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ABSTRACT

The objectives of the symposium were to define the problems of the child with spina bifida and to present practical means of management, using a multi-disciplinary team approach. Eight papers defining the problem cover the epidemiology of spina bifida, pathophysiology, musculoskeletal defects, incontinence of bladder and bowel, problems of function and of intellect, socio-economic problems, and the family. Six papers concern the following aspects of management: comprehensive management of the newborn, immediate repair, urologic management, management of hydrocephalus, mobility, and problem solving in management. Four papers discuss the older child in terms of educational programs, hip reconstruction, nursing, and adolescence. (KW)

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SYMPOSIUM
ON
SPINA BIFIDA

November, 1969

DEPARTMENT OF PHYSICAL MEDICINE
AND
REHABILITATION

BIRTH DEFECTS CENTER
DEPARTMENT OF PEDIATRICS

UNIVERSITY OF COLORADO
SCHOOL OF MEDICINE

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PROGRAM PARTICIPANTS

GUEST SPEAKERS

W. J. W. Sharrard, M.D., F.R.C.S.
Consulting Orthopedic Surgeon
Sheffield Royal Infirmary and Children's Hospital
Sheffield, England

Mrs. Margaret Hill, M.N.
Nursing Consultant, Division of Congenital Defects
Department of Pediatrics
University of Washington

Howard G. Rosen, A.C.S.W.
Regional Commissioner, Denver, Colorado
Social and Rehabilitation Service
Department of Health, Education and Welfare

Betty Green, Ph.D.
Professor, Department of Special Education
Colorado State College

UNIVERSITY OF COLORADO FACULTY

Alfred J. Defalco, M.D.
Instructor, Division of Urology

Claire A. Fishman, Ph.D.
Assistant Professor, Departments of Psychology and Physical Medicine and Rehabilitation

Jerome W. Gersten, M.D.
Professor and Chairman, Department of Physical Medicine and Rehabilitation

Harriet E. Gillette, M.D.
Professor, Department of Physical Medicine and Rehabilitation

Wolff M. Kirsch, M.D.
Associate Professor, Department of Surgery

Margaret McCall, M.S.W.
Social Worker, Birth Defects Center
Department of Pediatrics

James S. Miles, M.D.
Professor and Head, Division of Orthopedic Surgery

John H. Meier, Ph.D.
Director, John F. Kennedy Child Development Center

Ida I. Nakashima, M.D.
Assistant Professor, Department of Pediatrics

Albert F. Petrone, M.D.
Clinical Assistant Professor
Division of Urology

Arthur Robinson, M.D.
Professor and Acting Chairman, Department of Biophysics
Director, Colorado Birth Defects Center

Janet M. Stewart, M.D.
Assistant Professor, Department of Pediatrics
Assistant Director, Colorado Birth Defects Center

PREFACE

Recent advances in surgical techniques preserve function; diagnostic and therapeutic efforts measure and apply abilities; new techniques in the social sciences provide opportunities for productive living for children with birth defects. A coordinated team effort, with thorough understanding of the problems of each member is essential for successful management of the child with spina bifida.

The objectives of this symposium are:

1) To define the complex problems of the child with spina bifida, and those of his family.
2) To increase the awareness of physicians and allied health personnel as to the good potential of many children with spina bifida.
3) To present practical means of management of problems presented, using a multi-disciplinary team approach.

As members of the rehabilitation team direct their efforts toward a common goal, it may be anticipated that the potential for productive living, present in many persons with spina bifida, might be realized.

Harriet E. Gillette, M.D.
Professor,
Department of Physical Medicine and
Rehabilitation
University of Colorado Medical Center

GREETINGS

Dean David Talmage, M.D.

It certainly is a pleasure to welcome you to the opening session of this symposium on spina bifida. The business of the university is education; the particular business of the Medical Center is education of health care personnel. The present symposium, I believe, epitomizes the complexities of this role. To determine the truth and present it to the victims of human suffering honestly, but with compassion and with hope, to work toward improving our methods of preventing and correcting illness, to organize and train health care personnel, these are all extremely complex problems which this particular and unfortunate disease illustrates very well.

The University of Colorado Medical Center is pleased to sponsor this symposium. I know that you are going to have a successful and rewarding experience. I am pleased also to acknowledge the contribution and support of the National Foundation and the Department of Health, Education and Welfare through the training grant awarded to the Department of Physical Medicine and Rehabilitation.

THE PROBLEM DEFINED

THE ETIOLOGY AND EPIDEMIOLOGY OF MENINGOMYELOCELE AND ANENCEPHALY

Arthur Robinson, M.D.

In the western world meningomyelocele and anencephaly represent one of the commonest, and undoubtedly one of the most serious congenital malformations. In some countries it may be present in as much as 10% of live-births and up to 50% of still-births with gross malformations (1). As a chronic disease causing severe impairment of locomotion, meningomyelocele is second only to cerebral palsy in frequency and has the added hazard of being a frequent cause of mental retardation. These factors, the chronicity and the serious disability, including mental retardation, have placed physicians in ethical and therapeutic dilemmas which have been frequently debated in the medical literature (2) and will undoubtedly be discussed later in this symposium.

The neural tube is formed during the fourth week of gestation, and spina bifida results when closure of the neural groove which starts in the dorsal region is incomplete either cephalad or caudad. The two defects (meningomyelocele and anencephaly) therefore arise from the failure of a single process. That they have the same etiology (or etiologies) is further suggested by the frequency with which they both appear within the same family (3) and by their common epidemiological properties. It will be noted, however, that most of the larger epidemiologic studies have concentrated on the occurrence of anencephaly because it is obvious at birth, is invariably lethal and is readily ascertained. Meningomyelocele, on the other hand, may be under-reported since death certificates not infrequently list children with repaired spina bifida as "hydrocephalus."

Little is known about the etiology of these defects. Undoubtedly a heterogeneity of causes working cooperatively and differing in different individuals is involved. A variety of environmental agents have produced the condition in animals, and the evidence is persuasive that a similar situation exists in man, particularly in regard to the folic acid antagonists (Table 1, 4, a-c)

Table 1. Teratogenic Factors

Species	Agent
Hamster	X-ray
Mice	X-ray
Mice, Rats	Trypan Blue, Vitamin D, Salicylates
Man	Folic acid antagonists

Since the factors involved in the production of meningomyelocele are thus far undetermined, many have turned to epidemiological studies in the hope of thereby developing clues. The over-all incidence is roughly 1.5×10^{-3} births (5), but the most striking fact brought out by the various studies is the variability of the incidence in different regions of the world (Table 2). Why the incidence in Dublin should be 10 times greater than that in France is indeed a mystery.

Table 2. Occurrence of Anencephaly and Spina Bifida in Various Parts of the World

Dublin	5.9×10^{-3}	Ref.6
S. Wales	4.5×10^{-3}	7
Alexandria	3.79×10^{-3}	1
Birmingham (1940)	3.35×10^{-3}	8
Liverpool	2.8×10^{-3}	9
Buckinghamshire	2.8×10^{-3}	10
Rhode Island	2.5×10^{-3}	11
New England	2.5×10^{-3}	12
Rochester	2.4×10^{-3}	13
Bombay	1.9×10^{-3}	1
Mexico City	1.8×10^{-3}	1
Birmingham (1947)	1.6×10^{-3}	8
New York City	1.1×10^{-3}	14
Japan	0.8×10^{-3}	15
France	0.5×10^{-3}	16

In addition to these regional variations in incidence, yearly and seasonal variations have also been reported. For example, in Scotland and Dublin, there were peaks of incidence in 1940 and 1960 (5a) while, on the other hand, a significant drop occurred in Birmingham from 1940 to 1947, and in upper New York state between 1945 and 1959 (5b). In addition, whereas a fetus incurs a greater risk of anencephaly if born in Birmingham and Scotland during the winter, this does not seem to be true in Rhode Island. (3)

Of similar interest, but of unknown significance, are the statistics showing that affected babies are predominantly female (this is particularly true in anencephaly where the female/male ratio ranges from 1.9 to 4.2 (18) and that the incidence among negroes is less than among caucasians (19). However, the fact that first generation Irish in Boston had a higher incidence than the second and third generations (20) suggests that differences in the socio-economic status of the different populations studied may be important factors. The complexity of environmental agents which may be involved is further attested to by the consistent association that is present between meningomyelocele and city living (21).

There is little information to suggest that genetic factors make a significant contribution to etiology. Reports of twins showing concordance for neural tube defects have been few, and little, if any, difference has been demonstrated in concordance between monozygotic and dizygotic twins (1). However, despite all the evidence that non-genetic factors probably do predominate is the indubitable fact of increased family incidence with a tendency for neural tube defects to recur in sibships (3). Not infrequently, one sib will have spina bifida and the other anencephaly. Moreover, statistics from Egypt reveal an incidence of neural tube defects of 14.2×10^{-3} in the offspring of consanguineous marriages in contrast to a figure of 5.7×10^{-3} in the population at large (1).

In attempting to appraise the genetic component in etiology, particular attention should be paid to maternal influences. In South Wales the risk was doubled in the mother's sister's children (22), and the risk to maternal half-sibs (of affected offspring) is said to be as great as to full-sibs(22a). Perhaps something in the mother's genotype makes the intra-uterine environment less conductive to normal neural tube development or perhaps cytoplasmic inheritance is the determining factor.

In any case although genetic factors, possibly cytoplasmic, are operative, the evidence is clear that environmental factors are more important in this multifactorially determined disease.

Table 3. Factors Influencing Risk of Spina Bifida

Race	Caucasian
Geography	Urban vs. Rural
	Country - Ireland
Maternal age	Extremes
Season of birth	Winter
Socio-economic class	Lower
Sex	Female
Relative	Maternal
Teratogens	X-ray, anoxia, abortifacients

The epidemiological information here described has defined some predisposing factors which should be considered in genetic counseling (Table 3). Risk figures of course are completely empiric and must be considered averages (Table 4). The figures themselves may not appear to be too prohibitive if one does not consider the "stakes" as well as the "odds," the tragic nature of the disease making even a relatively small risk unacceptable to many people.

Table 4. Recurrence Risks for Neural Tube Defects (22,18)

One affected child	4-6%
Two affected children	10-12%
One affected child	
Risk of abortion	15-25%

Finally, what is the natural history of meningomyeloecle, and how effective is therapy? Both of these questions are difficult to answer and the latter, at least, will form the major part of this symposium. There have been notable differences in the various studies which have attempted to answer these questions, possibly due to differences in the kinds of cases selected. In general, one can say that

25% of affected newborns will die by the end of the first 24 hours, a figure which will probably be unaffected by active therapy. One study (23) has concluded that if untreated, only 10% will be alive by five years of age and that over half of these will be severely disabled.

On the other hand, Eckstein and McNab reported that with aggressive therapy, well over 50% of affected children will do well (24).

In conclusion, I hope I have succeeded in sketching the magnitude of the problem to be faced in the prevention and treatment of one of the most common and most disabling of congenital malformations.

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PATHOPHYSIOLOGY OF SPINA BIFIDA AND HYDROCEPHALUS

Wolff M. Kirsch, M.D.

Spina bifida can be a serious congenital malformation. The term is inclusive and signifies a midline defect in the bony structures which surround the spinal canal. This defect usually involves the posterior elements (laminar arches) but may rarely be situated anteriorly, involving the vertebral bodies. Since approximately 60% of all congenital malformations involve the nervous system or adjacent bony and soft tissue coverings, it is not surprising that spina bifida and associated problems (hydrocephalus, encephalocele) have stimulated the interest of a diverse group of investigators. As an aside, about 1% of all deaths occurring in the first year of life can be related to the presence of spina bifida (1).

THE FAUN--BERNINI

Figure 1.

The terminology used to describe the malformation relates to a continuum of anatomical defects which may be either harmless or incompatible with life. These catagories are arbitrary, but may be conveniently divided into spina bifida occulta, meningocele, meningomyelocele, myelocystocele, and myeloschisis. Spina bifida occulta, as the name implies, may be totally innocuous.

It is estimated that this disorder occurs with a frequency approaching 20% and can only be determined by radiological examination. The presence of this disorder may be signalled by an overlying cutaneous anomaly such as localized hairy overgrowth in the midline lumbosacral area or midline sinus tracts which may indicate an intraspinal epidermoid tumor. (2) An Italian sculptor, Bernini, who has never received the attention or acclamation he deserves was familiar with this cutaneous hypertrichosis, as were the ancient Romans, who regarded so-afflicted individuals as fauns. (Figure 1.) The term meningocele is reserved for a protrusion of meninges only, and meningomyelocele for the frequent and clinically troublesome presentation of neural tissues and meninges in a cystic sac. Myelocystocele is a rare association of central cystic cavitation of the cord attended by spina bifida.

The pathophysiology of these defects is really a study of teratology. There are many unsolved questions which have excited great controversy - not only with regard to etiology but also with respect to treatment. As stated by Warkany (3), observations of human cases have not answered the basic questions regarding etiology. Though a voluminous literature pertaining to the morphogenesis of spina bifida has accrued, particularly with the chick, (4), (5), relatively few studies of the problem have been conducted in mammals. The studies of Warkany and associates on the morphogenesis of trypan blue-induced meningomyelocele of the rat are of particular interest. (3) Though data derived from animal lesions can only be extrapolated to the human problem with caution (because of species differences), a workable experimental model is desirable to at least make an initial step in understanding the problem. Warkany's elegant studies in the rat have demonstrated a serial change in which a myeloschisis is transformed into a meningomyelocele. Thus a persistently open neural plate is considered to be the forerunner of the meningomyelocele, and this failure of differentiation occurs most frequently at the anterior or posterior neuropore - the last areas of the neural axis to close. Histologic serial sections of these lesions, studied between the twelfth and twenty-second day of gestation, have demonstrated that the exposed neural plate is ballooned dorsally by progressively collecting cerebrospinal fluid. This results in an arrangement of nerve roots into a row of four with the dorsal roots appended to the lateral wings of the exposed neural plate. The latter is subjected to both external trauma as well as a marginal blood supply and undergoes both degenerative and reparative changes. The final picture at the time of parturition is a central area medullo-vasculosa (6) as termed by Recklinghausen while at the periphery is an area epithelio-serosa comprised of pia and a thin layer of epidermis. In the human the entire sac may become epithelialized during the last half of pregnancy.

These anatomical studies, though of some value in recreating the anomaly by exposure to a teratogenic agent, tend to obscure other important factors which may be responsible for the process. It is important to distinguish two large groups of congenital malformations: 1.) those caused by arrested development during the period of organogenesis and 2.) those related to defects occurring after organogenesis in the period of further differentiation. In rat, for example, approximately 2/3 of the 22 day gestation period is involved in organogenesis - whereas in man only about 1/4 of the 269 days gestation period is concerned with organogenesis. A great deal can happen to the fetus during the remaining 3/4 time involved in gestation, particularly those problems which will be discussed in another section, e.g., hydrocephalus. Closure of the neural tube in the human occurs at about four weeks, and the assumption can be safely made that any responsible factor must have acted before this time to lead to a meningomyelocele. Thus knowledge of the embryological program can indicate or exclude certain suspected causal factors. This so-called "termination time" can only set a limit on the latest date at which a predicted injury or toxic agent might have acted to produce the anomaly, but cannot state the "determination time"- or precise moment when the stage was set for its development. More critical experiments are obviously needed to define this problem; namely, studies of the actual genetic control of the information required to program the growth and maturation of the exceptionally complex piece of machinery known as the nervous system. Since the timing of these developmental processes are relatively precisely known, and the importance of cellular events during the first trimester are crucial, attention has been paid in recent years to this period.

Helpful data has been obtained by autoradiographic techniques utilizing titrated thymidine. Several quite interesting facts have emerged from these studies: (a) Cells of the embryonic neural plate and neural tube are all of one cytological type (7) and organized as a pseudostratified epithelium. Nuclei engaged in the most extensive DNA synthesis are situated in the outer part of the cell, and after the synthetic stage they migrate toward the central canal and enter mitosis when adjacent to the central canal surface. The entire duration of synthesis, gap phase, and mitosis has been measured with the total cycle comprising about 8 hours. (b) During the generation cycle the cell will either migrate to form the primitive mantle, remain as primitive ependyma with attendent nuclear oscillations, or take an intermediate position between the mantle and central canal primitive ependyma. The controls which are operating to regulate this earliest stage of the differentiating process are virtually unknown and offer a potentially rewarding area for further study. Shortly after the specialization of the post-mitotic neuroblasts, the latter cells migrate secondarily to form regional specializations such as the intermediolateral cell column in the spinal cord. Lesions interposed at any time during this induction process could result in the final result of meningomyelocele. Only a rigorous quantitative examination of these effects will pinpoint the precise "determination time." (8)

Having examined the pathophysiology of spina bifida in the context of a failure in the early period of organogenesis, hydrocephalus (an associated anomaly) can be considered in the period following organogenesis. Current concepts regarding the physiology of the formation of the cerebrospinal fluid (CSF) have been recently reviewed. (9) It has been established that the rate of CSF formation is independent of hydrostatic pressure within the physiological range, and the rate varies considerably not only from species to species but from patient to patient. With regard to the site of its formation, it comes not only from the choroid plexus of the ventricles but in addition from unknown sources in the subarachnoid space. About 40% may originate from these sources. Absorption of CSF occurs over at least three different pathways -

only about 10% being directly absorbed into the dural sinuses, about 50% being diverted directly into the brain by indirect pathways. As the brain matures in utero, the resistance to CSF absorption decreases. The mechanism for this facilitated transependymal uptake is uncertain. One important factor is that the intraventricular CSF pressure is not fixed or constant, either during normal development or in hydrocephalus. The pressure increases with each pulse beat, primarily as a result of the choroid plexus filling with blood. Since these events are occurring in a closed cavity, and the brain is virtually incompressible, the pulsation forces out venous blood and CSF. The latter is displaced into the spinal canal. Any impairment of CSF flow will increase the ventricular pressure gradient and result in hydrocephalus. It is of interest that with a proximal blockade - i.e., aqueduct of Sylvius, ventricular dilatation occurs more rapidly than with more peripheral blockade - obstruction of subarachnoid pathways over the cerebral convexities. The management of these flow pressure problems will form the subject of the discussion regarding management of hydrocephalus.

To summarize, our understanding of the basic mechanisms responsible for spina bifida or hydrocephalus is imperfect. Any consideration of the pathophysiology of spina bifida must consider molecular events - or cell physiology, whereas hydrocephalus is an abnormal enlargement of the cerebral ventricles resulting from an increased pressure gradient between cerebral ventricles and brain. The latter, being a simpler concept to define and of recognized etiology, is thus more amenable to interventive therapy.

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MUSCULOSKELETAL DEFECTS

James S. Miles, M.D.

There are many ways to approach the clinical problems presented by the patients with myelodysplasia or spina bifida because the problems are so many, so varied, and cross so many specialty boundaries. Unfortunately, most articles approach the clinical problem in a very myopic fashion presenting only one small facet of the total clinical problem. The reader is then left to the difficult task of evaluating many differing clinical regimens, trying to co-ordinate many diverse treatment recommendations, and often reconciling conflicting opinions and suggestions. The real merit of a program such as is being presented in this course is to bring all of these varied thoughts together so that the participant in the course may evaluate all.

Most orthopedic texts treat the problem of myelodysplasia as a simple one in the management of the musculoskeletal defects. Lest we fault the orthopedist, let me observe that neurology texts and urology texts demonstrate similar myopia. It is also true that other medical fields such as social service and nursing will present similar biased views. Although I am charged with the responsibility of presenting the problems seen in the musculoskeletal system, let me state that such are but a part of the over-all problem presented by these patients, and that I wholeheartedly endorse the concept of total care of the patient, and that I insist that the patient must first and foremost have a doctor capable of seeing the totality of problems. I do not care what specialty designation that doctor has; he must be a doctor first and a specialist second.

To meet my obligation to present the subject of musculoskeletal defects, let me first approach it as a simple tissue problem. I shall comment first upon the osseous problems, then progress to the joint problems, and finally call upon Dr. Sharrard to add information on the muscular defects and problems. We must realize that all are related, and that it may be impossible to separate them.

The first and most obvious problem is the defect in the vertebral column. Most commonly the vertebral defects are in the posterior elements of the vertebrae, although some patients may present some extensive defects involving the entire vertebra or vertebrae. The posterior elements normally develop in the mesodermal tissues which lie posterior to the neural cord after closure of the neural tube. The posterior vertebral elements consist of paired, bilateral bony arches which should join in the mid-line posteriorly after closure of the neural tube. These osseous elements are the pedicles, the transverse processes, the articular facet processes (both superior and inferior), the laminae, and the spinal process. If the neural tube does not close, the mesodermal osseous elements do not unite, and a radiologically visible "spina bifida" results. The dysplastic development may be minimal or it may be very extensive. Thus there may be simple spina bifida occulta in which case the posterior elements of the vertebra come very close to each other, but do not quite unite by osseous tissue. The roentgenogram may indicate a simple gap of osseous tissue of a few millimeters or so. One should be careful in his interpretation of the roentgenogram, for although the film may indicate a gap of osseous tissue, this must not be interpreted as an absence of tissue in that area. In most instances a radiolucent tissue is present there such as cartilage or fibro-cartilage or strong fibrous tissue, and the resultant vertebral arch is almost as strong as it would be if there were a complete osseous arch. Or there may be complete loss of the posterior elements in an osseous form with no suitable structural tissue substituted with resultant loss of stability of the vertebral column. In addition, as has been mentioned, the vertebral bodies anteriorly may be a part of the dysplasia demonstrating all sorts of anomalies from simple deformity to complete absence.

Obviously one must consider the vertebral column from a standpoint of function, and that function for the vertebra must be considered to be stability, structural strength, and protection of the cord. If the osseous loss or dysplasia is extensive, there may be weakness of the vertebral column from a structural standpoint. Such weakness may not be apparent as long as the infant is in the horizontal position, but may be quite obvious as the child becomes erect. There may be kyphosis or scoliosis of significant degree. Other tissues such as cartilage and muscle may try to compensate for the loss of osseous tissue, but these may also be involved in the dysplasia, and therefore unable to compensate adequately. The loss of vertebral stability will vary from minor with the production of no symptoms (or a mild, intermittent low-back pain) to serious deforming and disabling spinal curvatures--gibbus formation, kyphosis and scoliosis. Some of the vertebral deformities may be progressive as the increasing stresses of weight bearing, activity and muscle weaknesses or paralyses are added.

Fortunately, the minor osseous defects rarely need treatment. Other tissues usually provide enough stability for the activity to be anticipated in this patient with somewhat limited physical potential. Moderately severe osseous defects likewise rarely require treatment, for the other physical impairments of the patient may limit the habilitation potential of the patient, and the osseous defect itself may not be the limiting factor. Unfortunately, some of the very serious osseous defects in themselves may be disabling. The severe kyphosis or scoliosis may affect the patient much more than cosmetically, producing asymmetric weight bearing, bed sores, or a deformity incapable of bracing. These severe osseous defects and deformities require a great deal of thought, and an evaluation of the total capability of the patient so that a meaningful reconstructive operation can be planned. Such an operation may well include the resection of hemivertebrae, the production of an anterior inter-body vertebral fusion, or simple vertebral resection to produce a flail, non-deformed vertebral column.

Other osseous defects may be present and are a part of the total problem of faulty development in these patients. Fortunately extremity phocomelias and absences are very rare. If present, they may present the tremendous variabilities of the extremity defects seen in patients without myelodysplasia. However, these patients usually have sensory problems as well, and are a more difficult problem for surgery or prosthetic fitting because of such. The usual orthopedic reconstructive surgical and prosthetic procedures should be considered, and will not be discussed here.

On the contrary, joint problems are extremely common in the patients with myelodysplasia. The joint deformities are almost exclusively in the lower extremities, and may include one or all of the joints of one or both of the lower extremities. The deformities may be of contracture or of subluxation and dislocation. In some respects the deformities of the joints may appear to be quite similar to those which occur as poliomyelitis residuals, or in those patients with cerebral palsy. However the only real similarity is in the positions assumed. The deformities of all of these patients are due to muscle imbalances about the joints and the effects of gravity. In patients with the residuals of poliomyelitis and those with myelodysplasia, the muscle imbalance is due to varying degrees of paralysis. The patients with cerebral palsy have variable and asymmetric degrees of spasticity with resultant joint deformities. One major difference is apparent in the patients with myelodysplasia--usually there is a sensory deficit as well as a muscle deficit. Such a sensory loss is not present in the patients with the residuals of poliomyelitis. The sensory loss in the patients with myelodysplasia makes for surgical and bracing problems.

Another common problem of these patients with myelodysplasia is dislocation of the hip. The dislocation is secondary to muscle imbalance. It is accompanied by deformities of the proximal femur and the acetabulum. Such deformities are visible in roentgenograms early in the life of the patient, and should require treatment by the physician to prevent their progression to complete dislocation of the hip. Adequate treatment often consists of muscle transfer or reconstructive procedures about the hip. These may include osteotomy of the femur, the pelvis, or both.

I would like at this time to ask Dr. Sharrard to discuss these muscle imbalances as they relate to the musculoskeletal defects.

MUSCULOSKELETAL DEFECTS IN SPINA BIFIDA

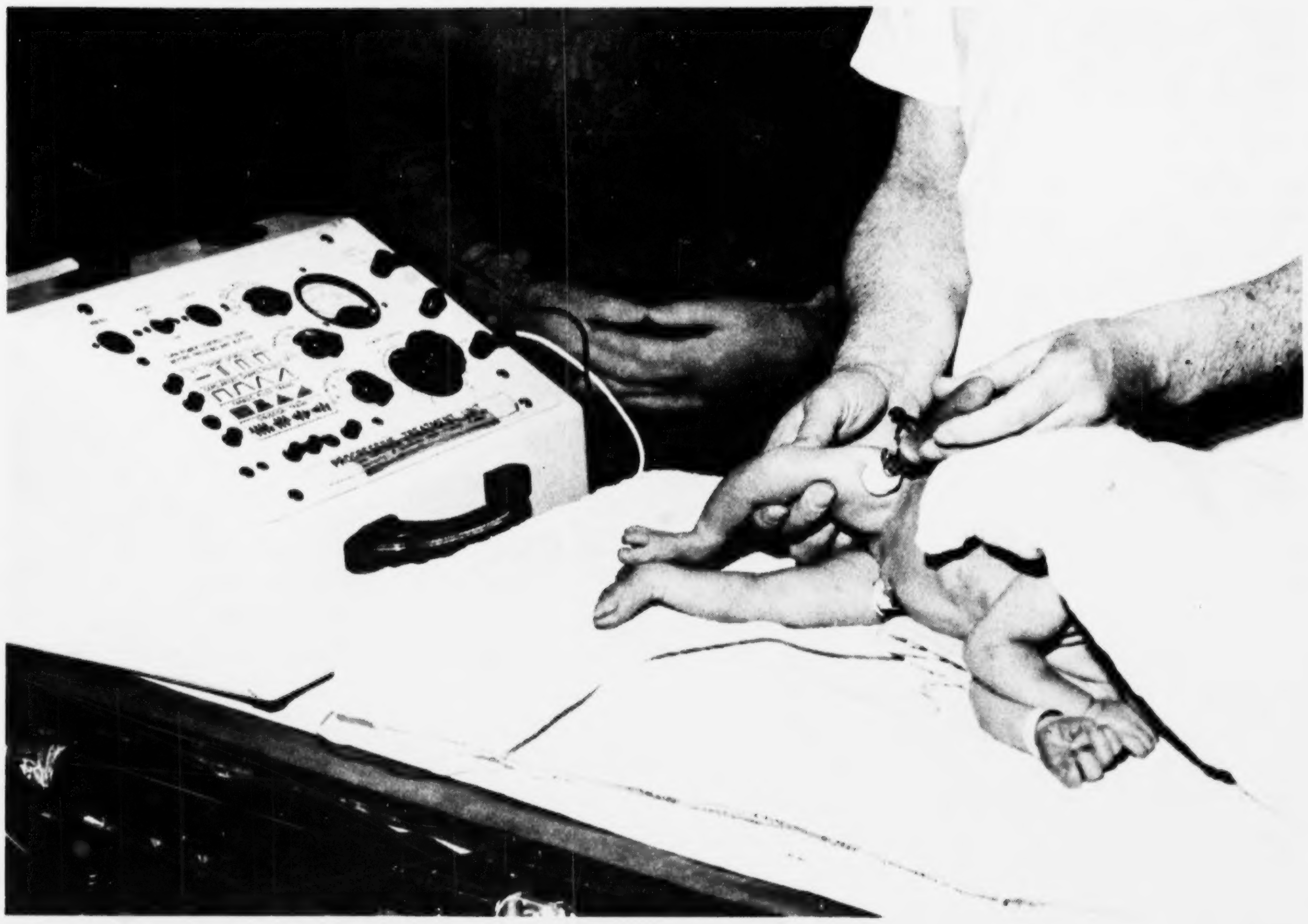
W.J.W. Sharrard, M.D., F.R.C.S.

In children with spina bifida, deformities may be present at birth or may develop after birth. Some of the congenital defects have the appearance of a coincident deformity of spina bifida and talipes equino varus. When one considers all the deformities that may exist, then appearances are much more like the type of deformity that tends to occur in poliomyelitis and other conditions which are the result of flaccid paralysis.

This observation led me to make an extensive study into what paralysis these children have at birth and to try to make some inferences as to what paralysis they may have had in utero. All investigations by electrical stimulation, electromyography, biopsy and necropsy studies suggest that the deformities in the lower limbs that are present at birth are secondary to the effects of the paralysis which is present in utero.

I will ask those of the ladies who have had babies, and husbands whose wives have had babies to remember that infants kick in utero at quite an early stage, and one could suggest the thesis that if some muscles are working and if some are not when the baby is starting to kick in utero, that the working muscles would be likely to pull the foot, hip or knee out of shape if the antagonists are not there to pull back. The situation that makes the analysis difficult is that when the babies are born they have just been precipitated through an unpleasant journey along the vagina with the spinal cord on the surface, and in addition the spinal lesion may have been compressed by forceps during delivery. It is no surprise therefore that quite a proportion of these children, when they arrive in the world, have no muscles working in their lower limbs at all.

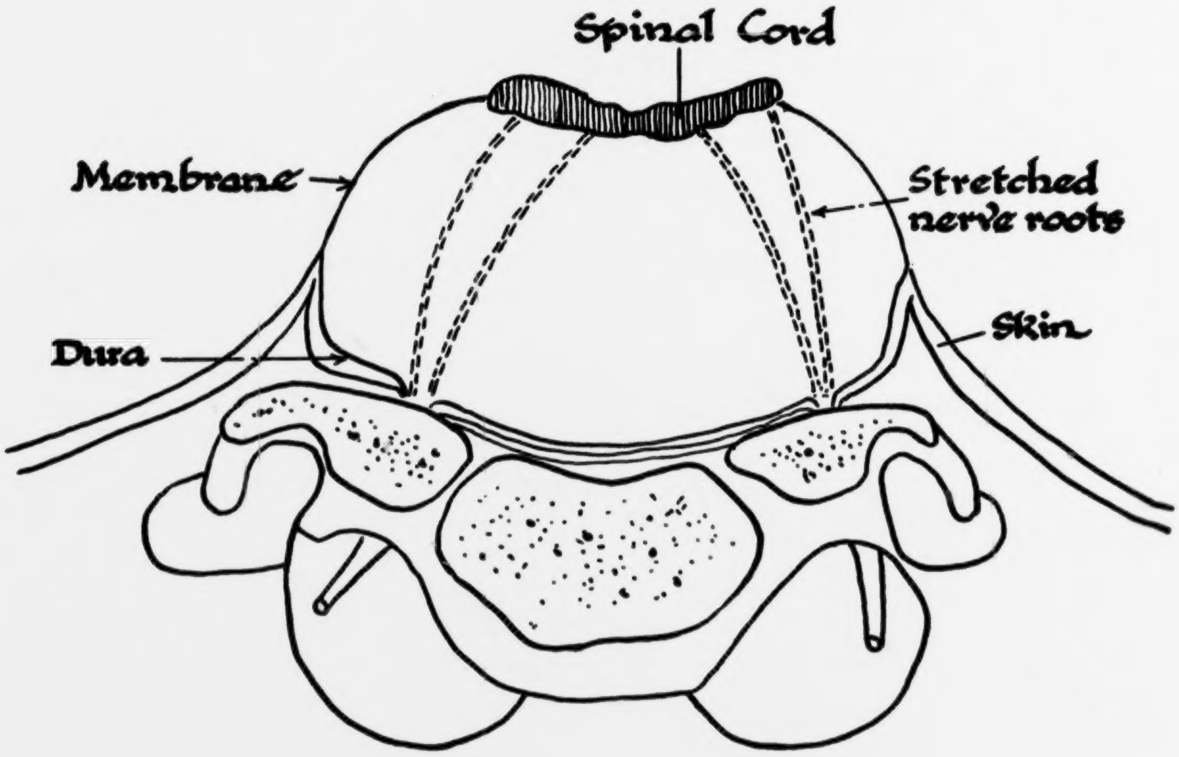
We have attempted to determine the number of functioning nerve cells in the neural plaque and have found large numbers of intact nerve cells, anterior horn cells in particular, but distributed in such a way as to make analysis very difficult. However, by counting the proportion of normal-appearing nerve cells from the plaque in relation to those found in the normal we have found that in 20% of children with meningomyelocele there is a normal number of anterior horn cells with normal appearance and we think these are potentially able to function. What is not possible to estimate from such an analysis is how many of these may be connected proximally in the normal way. In the remaining 80% there are varying degrees of deficit and in many instances these cells correspond to innervation from the lumbar segments. For instance, there can be normal innervation from the first four lumbar segments with absence of cells, nerve roots and spinal cord below this level. When a child has this distribution of neural loss there will be action in the tibialis anterior and posterior and perhaps in tensor fasciae latae.



Percutaneous faradic stimulation in neo-natal spina bifida

Figure 1.

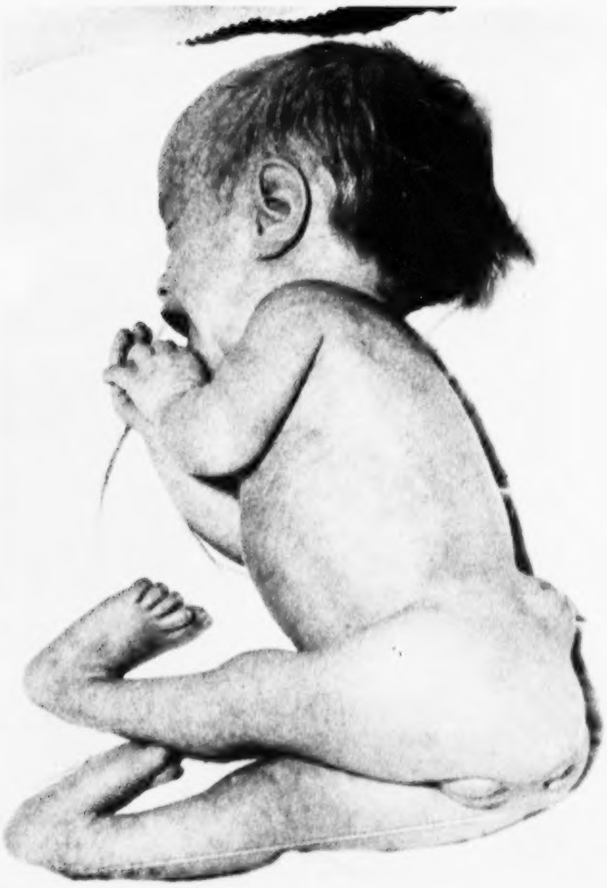
We have found that the paralysis which we often see in a child two or three hours old is only a partial paralysis and, if given correct treatment, it can recover. The problem then is to find out how much of the paralysis in the legs is true and how much is temporary. To sort this out we do a direct stimulation with faradism to the lower limb muscles on the first day of life before the lesion is closed (Figure 1) and when doing this we have found that the deformity correlates almost exactly with the results of faradic stimulation. This results is some typical patterns of deformity.



The pathological anatomy of meningomyelocele
(From Baraffaldi and Divano)

Figure 2.

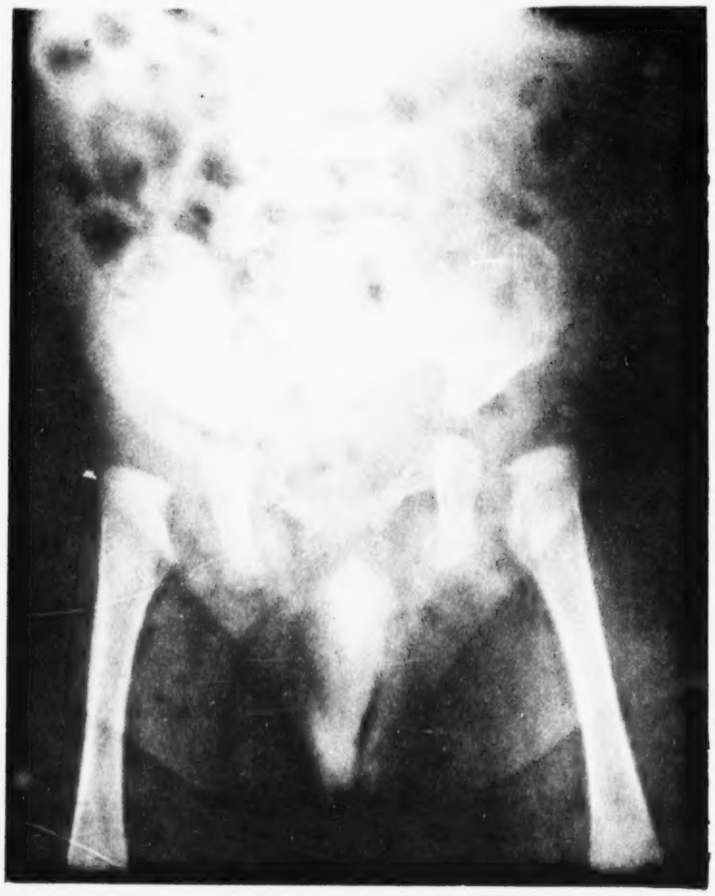
A diagram of the features of meningomyelocele shows the pathological anatomy (Figure 2). The spinal cord in its lower part opens out into a plaque which is exposed on the surface and which may close up again distal to the lesion. The diagram shows the plaque to be bulging away from the surface but this feature does not usually develop until 24 to 48 hours later. The nerve roots which pass across from the plaque forward to the vertebrae become stretched; when we have taken sections of these nerve roots we have often found that this is a traction lesion. In addition to this the opened-out cord may itself be abnormal and contribute to the paralysis.



Lower limb deformities in paralysis below L_4.

Figure 3.

If we suppose that these muscles are working in utero and are causing deformity in the limbs we shall expect to find a baby who has flexed adducted hips, extended knees and calcaneo-varus feet (Figure 3). These deformities are rigid. The hips cannot be extended or abducted manually even at birth. Associated with this the hip is invariably dislocated and the knees are in recurvatum. There is complete paralysis of the glutei, and muscles that might flex the knees such as the gracilis and sartorius dislocate to the extensor surface and become extensors of the knee. The quadriceps is tight and there is not only recurvatum but limitation of flexion so that the knee will not flex more than 20 degrees because the quadriceps is short. The foot deformity is not a talipes equino-varus but a talipes calcaneo-varus. The inversion is very gross, often 90 degrees or more together with 90 degrees of calcaneus. The x-ray of the hips at birth shows that they are completely dislocated, but you should note that the acetabulum is well formed. (Figure 4.)



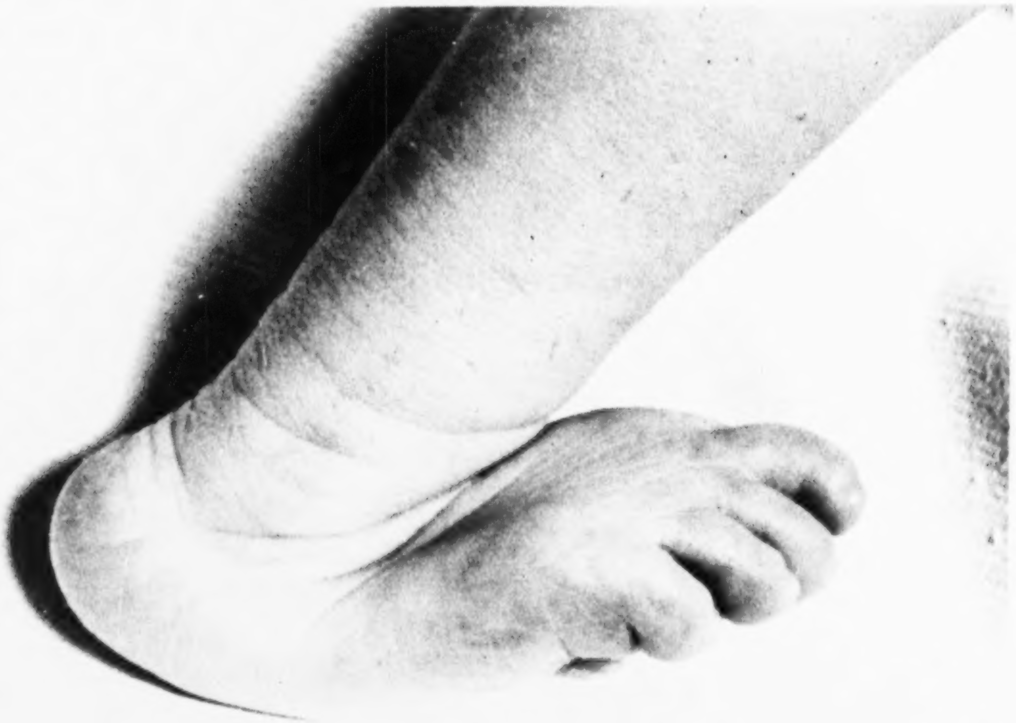
Radiograph of congenital paralytic dislocation of the hip

Figure 4.

In considering correction of these deformities sensory loss is very important. At birth some babies may even show a pressure sore on the sole of the foot. Pressure sores can even be caused by the uterus and this suggests that the application of Dennis-Brown splints or any such forcible mechanisms would be certain to cause yet more pressure sores. I think that it should also be noted that the hip is laterally rotated, a feature that is not present in ordinary congenital dislocation of the hip. This arises because the psoas in addition to being a flexor and adductor is a very strong lateral rotator. The thigh is so much laterally rotated that the greater trochanter is almost lying in the acetabulum.

If there is innervation down to the 5th lumbar segment, there are a number of muscles that are acting, in addition to those already mentioned. These include extensors of the toes, peronei, gluteus medius and some of the hamstrings, so that the deformity is now different. The hips are a little flexed and adducted but not grossly so. The knee is more normal but the feet are dorsiflexed because all the dorsiflexors are present and there are no plantar flexors. On x-ray the hip is not dislocated but it may be subluxated. The hip will continue to become more subluxated and eventually will dislocate, usually at about six months, but sometimes as late as the second year.

With innervation down to the 1st sacral segment there is paresis of the calf and gluteus maximus and of the long toe flexors and paralysis of the intrinsic muscles of the foot. In this situation when the strong dorsiflexors act they pull up the forefoot whereas normally if the intrinsic muscles were present they would transmit this pull to the os calcis. Since the intrinsics are paralysed the foot breaks in the middle, the talar and navicular joint dislocates and the result is a so-called vertical talus (Figure 5). Radiographs show that the os calcis is in equino-valgus. The talus which has no muscle attachments is flexed downward so that its head faces into the sole of the foot. This vertical talus deformity is occasionally seen in the absence of spina bifida, but it is not at all uncommon as an associated deformity with this condition.



Paralytic vertical talus

Figure 5.

Finally with minimal paralysis of the intrinsics there is a slight deformity confined to the foot which tends to be in varus with clawing of the toes, a picture that is often seen in spina bifida occulta.

Not all deformities are quite as straightforward as the ones I have described, particularly as the child grows and upper motor neuron innervation starts to show. There may be a complex picture of lower and upper motor neuron loss, especially if there has been secondary paralysis superimposed on that which was already present at birth. For example, one can have a child with normal innervation down to the 5th lumbar segment with paralysis below this level but with spasticity due to interruption of descending tracts in the lumbar segments. In this situation the result is some of the features that present in cerebal palsy with a spastic hip and knee together with some of the features of the lower motor neuron loss such as calcaneus feet. In a spina bifida population one can find almost any kind of deformity that exists.

The degree of paralysis and deformity does not necessarily relate to the size of the spinal defect. It is possible to have a child with an extensive spina bifida from the lower thoracic region to the sacrum and yet have normal or almost completely normal lower limbs. By contrast one might have a very small lesion in the lumbo-sacral region with quite extensive neural deficit.

In assessment both at birth and later, it must be determined what muscles are present and acting voluntarily, which muscles are reflex and which are completely paralysed. There is one particular variety of muscle deficit that has not been described before and which tends to occur in all these children. This is a situation in which the muscle appears to be completely paralysed without voluntary movement or reflex activity, but yet recurrent deformity develops particularly in equinus in spite of all efforts by parents and physiotherapists to prevent recurrence of deformity. In this situation if we divide the tendo-Achilles and correct the deformity it recovers very rapidly. Electrical stimulation shows that this muscle is in fact still active and this can be confirmed at operation. These muscles are ones which have retained motor innervation but have lost their sensory innervation, a situation which is quite peculiar to children with spina bifida.

When seeing these deformities at birth the natural instinct of any orthopedic surgeon is to feel that he ought to do something about it. With ordinary congenital dislocation of the hip and talipes equino-varus it is normal to attempt splintage immediately or at any rate during the first week of life. In spina bifida immediate treatment of this sort is not indicated for many reasons. The application of splints and plasters embarrasses the activities of one's neurosurgical colleagues in their treatment of the back and of the head. They are not able to care properly for the spine and cannot put up intraveneous transfusions. Another reason for not doing anything is that conservative treatment will only succeed if the muscles are still working and any attempt to force correction will inevitably cause pressure sores. Yet another reason is that it is very important that the child should at some early age return to his mother for a while; if possible we like them to get back home within six weeks. We tell the parents not to worry about the leg deformities and that we will do something about the deformities later on. A baby at this age needs care from his mother and at this stage, if he is left away from his mother, it may well be that such a child, especially one with deformities, will be rejected if he is kept in hospital. By allowing the babies to return home we have had a very low rejection rate indeed.

Orthopedic treatment can be started any time after three months, but in practice it usually works out that orthopedic treatment is started between six and nine months when the tissues are large enough to operate on and one can make a proper estimate of the paralysis and deformity that is present.

DIAGNOSIS AND MANAGEMENT OF THE NEUROGENIC BLADDER

Albert Petrone, M.D.

The primary objective in the diagnosis and management of the neurogenic bladder is the preservation of renal function. It has been repeatedly demonstrated that the deranged hydrodynamics of the bladder lead to persistent urinary infection and upper urinary tract destruction. It therefore becomes essential to make a correct diagnosis of neuromuscular dysfunction and to institute the proper measures to prevent the destruction of renal parenchyma.

Rose, in a study of 80 patients with meningomyelocele, found that nearly all patients who survived the first two years of life had a damaged urinary tract, 77% had persistent or recurrent bacteriuria, 40% had decreased renal function and 75% had uretero-vesical reflux. Only one patient had urinary control and had survived more than two years without urinary damage. (1)

Chapman and Associates in a study of meningomyelocele up to the age of four, showed that 73% had positive urinary cultures, 100% had abnormal cystograms, 25% had uretero-vesical reflux and 30% had abnormal intravenous pyelograms. (2)

The old classification of spastic bladder versus flaccid bladder represented by the inherent behavior of the bladder is not sufficient. Rather one should attempt to classify neurogenic disease of the bladder on the location of the neurological lesion and its involvement of motor or sensory tracts.

Based upon anatomical and physiological involvement of the nervous system, neurogenic bladder can be classified into the following types:

1) uninhibited neurogenic bladder
2) sensory paralytic bladder
3) motor paralytic bladder
4) reflex neurogenic bladder
5) autonomous neurogenic bladder (3)

ANATOMY AND PHYSIOLOGY:

A knowledge of the physiological properties of the bladder musculature and the neurophysiology relating to the control of this viscus is essential. The bladder is made up of smooth muscle which possesses the inherent property of tonicity. It has been demonstrated that the bladder will maintain its normal tonicity or ability to resist stretching even when there is an interruption of all its extrinsic nerves.

The bladder musculature becomes atonic or flaccid only when it has been stretched over a period of time to the point of fatigue. Thus, any condition which results in over distention of the bladder will result in a flaccid atonic bladder.

Another inherent property of the bladder musculature is the ability to maintain a constant intervesical pressure with increasing volume of urine. This power of accommodation is also completely independent of the central nervous system.

The nervous control of the bladder consists of a simple spinal reflex and a suprasegmental conditioned reflex. The afferent and efferent fibers of the spinal reflex arc are mediated through the pelvic nerves; the motor fibers are part of the parasympathetic nervous system S2,3 and 4. The normal stimuli for the reflex of micturition is filling of the bladder until proprioceptive impulses have reached a sufficient threshold to produce a discharge of the motor neuron, resulting in a voiding contraction of the bladder. In normal individuals the discharge of the motor neuron is under the control of the cortical regulatory tracts, the efferent portion of the suprasegmental reflex. The response of the upper motor neurons of the motor cortex and corticospinal tracts is influenced by the sensory impulses arising from the bladder and transmitted via the lateral spinothalmic tracts and the fasciculus gracilis.

NORMAL BLADDER

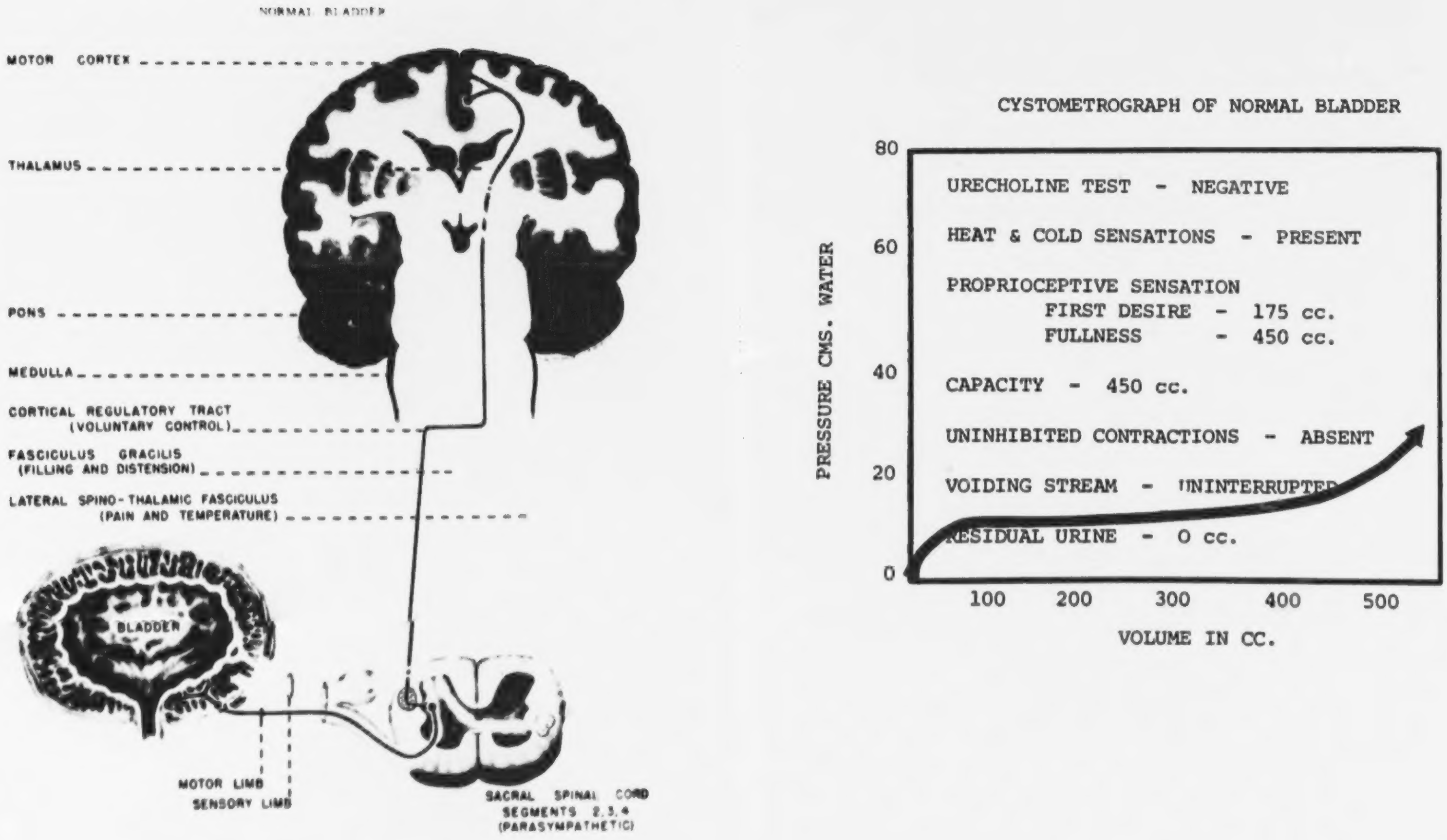

NORMAL BLADDER
MOTOR CORTEX
THALAMUS
PONS
MEDULLA
CORTICAL REGULATORY TRACT (VOLUNTARY CONTROL)
FASCICULUS GRACILIS (FILLING AND DISTENSION)
LATERAL SPINO-THALAMIC FASCICULUS (PAIN AND TEMPERATURE)
BLADDER
MOTOR LIMB
SENSORY LIMB
SACRAL SPINAL CORD SEGMENTS 2,3,4 (PARASYMPATHETIC)
CYSTOMETROGRAPH OF NORMAL BLADDER
URECHOLINE TEST - NEGATIVE
HEAT & COLD SENSATIONS - PRESENT
PROPRIOCEPTIVE SENSATION
FIRST DESIRE - 175 cc.
FULLNESS - 450 cc.
CAPACITY - 450 cc.
UNINHIBITED CONTRACTIONS - ABSENT
VOIDING STREAM - UNINTERRUPTED
RESIDUAL URINE - 0 cc.
PRESSURE CMS. WATER
0
20
40
60
80
100
200
300
400
500
VOLUME IN CC.

FIGURE 1.

In the normal individual, the function of the urinary bladder is controlled by the suprasegmental conditioned reflex mechanism. Interruption in any of the pathways that mediate this controlling mechanism of micturition results in abnormal vesical function that derives from the pathway affected.

In the normal bladder cystometrogram the pressure remains constant with the bladder filling until the time of voiding. Hot and cold sensation is present. The first desire to void is at approximately 175 cc. and the bladder capacity is 450 cc. The injection of 2.5 mg. of Urecholine subcutaneously does not raise the intervesical pressure greater than 15 cm. of water. In the denervated bladder the intervesical pressure rise is greater than 15 cm. of water. This results from the fact that the ganglionic synapses and neuromuscluar junctions of the denervated end organ are hypersensitive to cholinergic agents. (4)

UNINHIBITED NEUROGENIC BLADDER

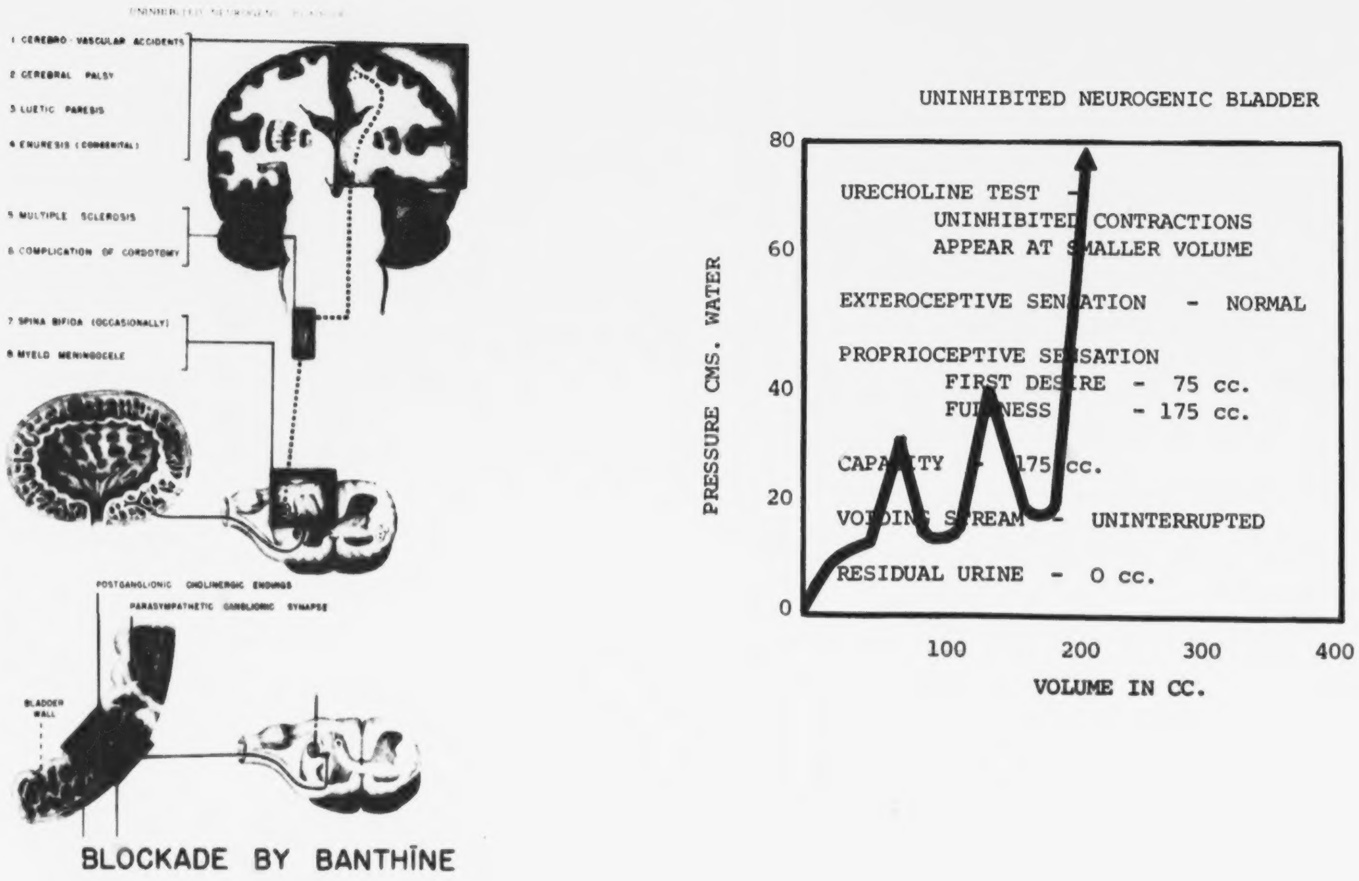

UNINHIBITED NEUROGENIC BLADDER
1 CEREBRO - VASCULAR ACCIDENTS
2 CEREBRAL PALSY
3 LUETIC PARESIS
4 ENURESIS (CONGENITAL)
5 MULTIPLE SCLEROSIS
6 COMPLICATION OF CORDOTOMY
7 SPINA BIFIDA (OCCASIONALLY)
8 MYELO MENINGOCELE
POSTGANGLIONIC CHOLINERGIC ENDINGS
PARASYMPATHETIC GANGLIONIC SYNAPSE
BLADDER WALL
BLOCKADE BY BANTHINE
UNINHIBITED NEUROGENIC BLADDER
URECHOLINE TEST -
UNINHIBITED CONTRACTIONS
APPEAR AT SMALLER VOLUME
EXTEROCEPTIVE SENSATION - NORMAL
PROPRIOCEPTIVE SENSATION
FIRST DESIRE - 75 cc.
FULLNESS - 175 cc.
CAPACITY - 175 cc.
VOIDING STREAM - UNINTERRUPTED
RESIDUAL URINE - 0 cc.
PRESSURE CMS. WATER
0
20
40
60
80
100
200
300
400
VOLUME IN CC.

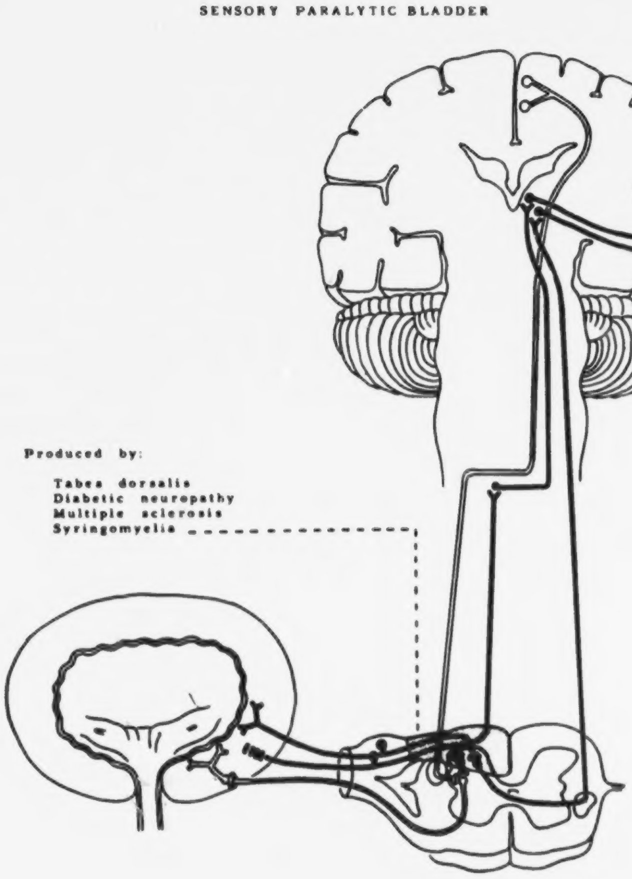
FIGURE 2.

The uninhibited neurogenic bladder resembles that of the infant. The patient is aware of the desire to void as the bladder fills, but is unable to inhibit the desire to void and voiding occurs. This condition presents clinically with urgency, frequency, nocturia, enuresis and in severe cases with urge incontinence. The lesion is an upper neuron lesion involving the cortical regulatory tracts or the cerebral cortex. It is seen in the newborn child, the hydrocephalic infant, in patients with cerebral vascular accidents, in multiple sclerosis, brain tumors, cerebral arteriosclerosis and occasionally spina bifida and meningomyelocele.

The cystometrogram reveals hypertonicity and uninhibited bladder contractions. Bladder sensation is normal. The bulbocavernous reflex and saddle sensation are present. The bladder capacity is reduced and residual urine is absent. The injection of Urecholine results in uninhibited contraction appearing at smaller volumes.

Management of this type of neurogenic bladder depends upon the ability to decrease the frequency and magnitude of the uninhibited contraction. This is accomplished by the use of anticholinergic drugs such as ProBanthine.

SENSORY PARALYTIC BLADDER



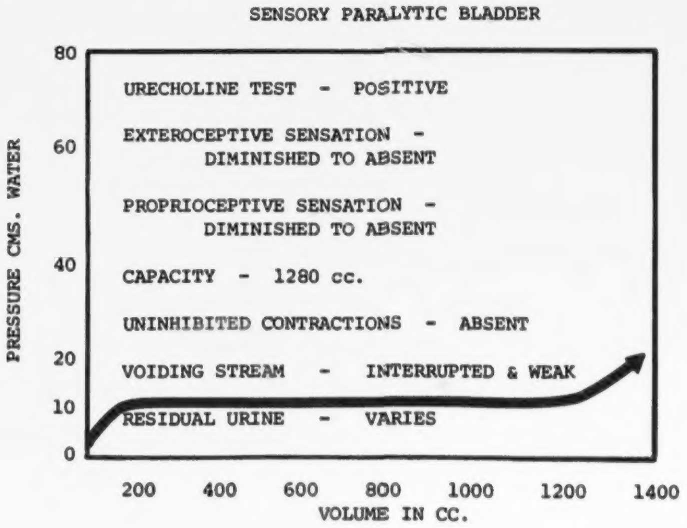


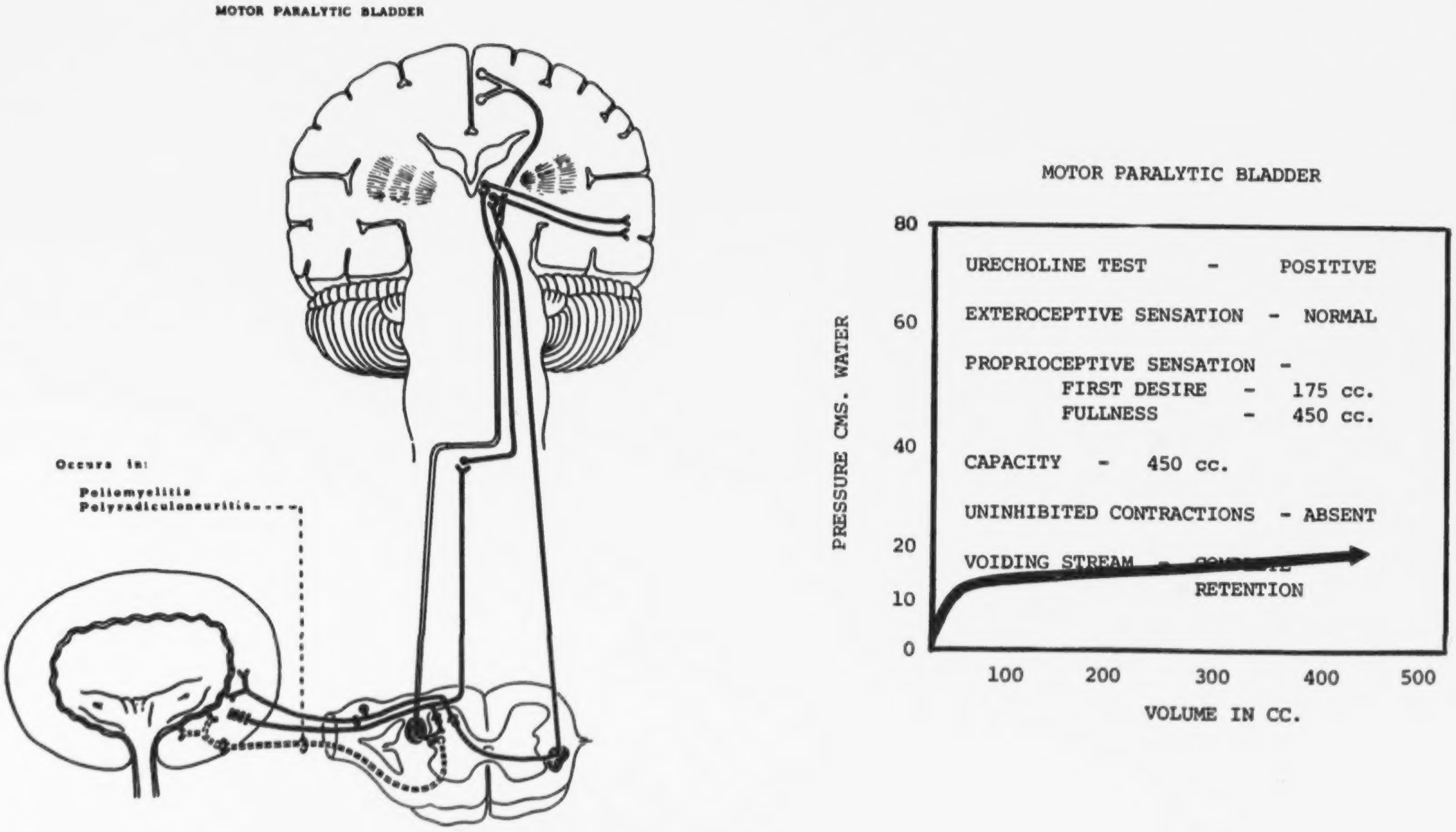
FIGURE 3.

The sensory paralytic bladder frequently presents clinically with distension or with overflow incontinence. The patient is not aware of the bladder filling and does not receive any stimuli to void from the filled bladder. This lesion involves the lateral spinothalmic tracts and is seen frequently in diabetes mellitus, tabes dorsalis and multiple sclerosis.

The cystometrogram displays a hypotonic, large capacity bladder without uninhibited contractions. The residual urine is usually increased. Bladder sensation to hot, cold and distension is absent or impaired. Saddle sensation is variable but the bulbocavernous reflex is present. The Urecholine test is positive, that is, there is an increase in intervesical pressure greater than 15 cm. of water.

This condition is probably the easiest variety to manage provided the bladder has not been over distended for so long that the detrusor muscle cannot recover. Management depends upon having the patient void at regular intervals by the clock, rather than waiting for a stimulus which will not appear.

MOTOR PARALYTIC BLADDER


MOTOR PARALYTIC BLADDER
Occurs in:
Poliomyelitis
Polyradiculoneuritis
MOTOR PARALYTIC BLADDER
PRESSURE CMS. WATER
0
10
20
40
60
80
100
200
300
400
500
VOLUME IN CC.

URECHOLINE TEST - POSITIVE

EXTEROCEPTIVE SENSATION - NORMAL

PROPRIOCEPTIVE SENSATION -
FIRST DESIRE - 175 cc.
FULLNESS - 450 cc.

CAPACITY - 450 cc.

UNINHIBITED CONTRACTIONS - ABSENT

VOIDING STREAM - COMPLETE RETENTION

FIGURE 4.

The motor paralytic bladder is extremely rare. It appears clinically as distension and occasionally with overflow incontinence. It must be differentiated from anatomically obstructing lesions such as prostatism. The neurological lesion involves the motor outflow from sacral segments S2, 3 and 4. It is seen in lesions of the cauda equina, poliomyelitis and peripheral neuropathy.

The cystometrogram shows a hypotonic bladder without uninhibited contractions. Bladder sensation is normal. Saddle sensation is normal but the bulbocavernous reflex is absent. The Urecholine response is markedly elevated, sometimes raising the intervesical pressure to 80 cm. of water or more.

Its management requires some form of bladder drainage or urinary diversion. Partial paralysis may be improved by the use of cholinergic drugs such as Urecholine. Fortunately, the recovery from the dysfunction of poliomyelitis and peripheral neuropathy is usually prompt.

REFLEX NEUROGENIC BLADDER

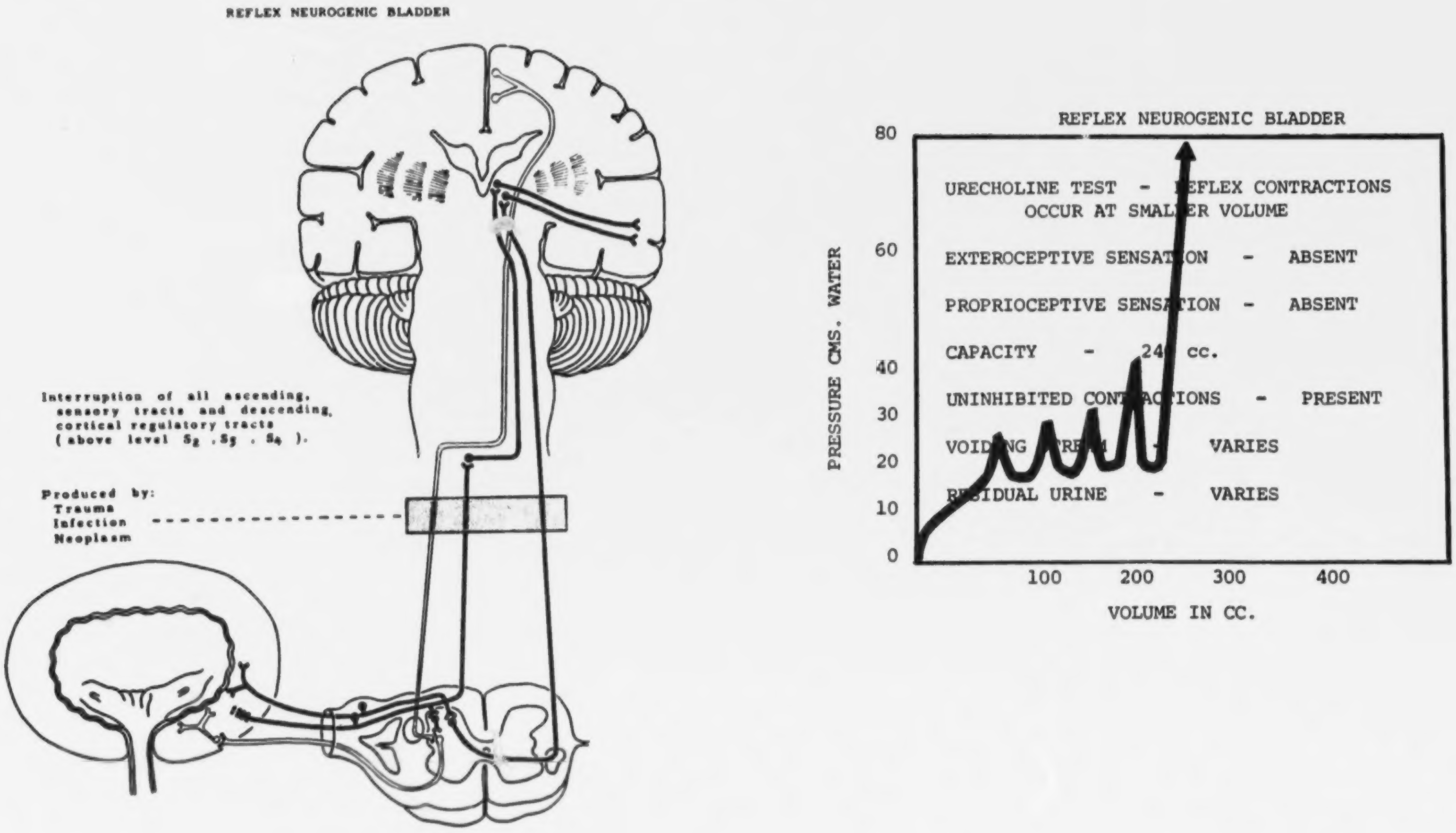

REFLEX NEUROGENIC BLADDER
Interruption of all ascending, sensory tracts and descending, cortical regulatory tracts (above level S_2 , S_3 , S_4).
Produced by:
Trauma
Infection
Neoplasm
REFLEX NEUROGENIC BLADDER
URECHOLINE TEST - EFLEX CONTRACTIONS
OCCUR AT SMAL ER VOLUME
EXTEROCEPTIVE SENSAT ON - ABSENT
PROPRIOCEPTIVE SENS TION - ABSENT
CAPACITY - 24 cc.
UNINHIBITED CON AC IONS - PRESENT
VOID NG TRE - VARIES
R IDUAL URINE - VARIES
PRESSURE CMS. WATER
80
60
40
30
20
10
0
100
200
300
400
VOLUME IN CC.

FIGURE 5.

The reflex neurogenic bladder results from transection of the spinal cord above the sacral segment leaving the reflex arc intact. It is seen in transections of the spinal cord, metastatic neoplasms of the spinal cord, multiple sclerosis, pernicious anemia and meningomyeloceles above T12.

Clinically, the patient presents with incontinence; voiding is precipitous, involuntary, frequent and interrupted. The bladder does not empty completely with involuntary voiding and residual urine is present. In addition, the patient's sensation of bladder filling is absent or impaired; nevertheless, some sympathetic responses such as sweating, abdominal fullness, nervousness and vomiting may appear.

The cystometrogram demonstrates absent sensation, bladder hypertonicity with uninhibited contraction, often insufficient in emptying the bladder, thus leaving a large residual urine. Saddle sensation is impaired or absent and the bulbocavernous reflex is hyperactive. The Urecholine test causes reflex contraction to occur at a smaller volume.

Management of the reflex neurogenic bladder requires continuous catheter drainage until the patient has stabilized. At this point the catheter can be removed and suprapubic pressure and Urecholine is used. Some will be able to void and continue in this manner. The majority will void reflexly and will be unable to empty their bladder. They will have uninhibited contraction and wet themselves. When there is evidence of upper urinary tract deterioration, persistent lower urinary tract infection or obstruction of the ureter by a hypertrophied bladder wall, permanent urinary diversion must be carried out.

AUTONOMOUS NEUROGENIC BLADDER

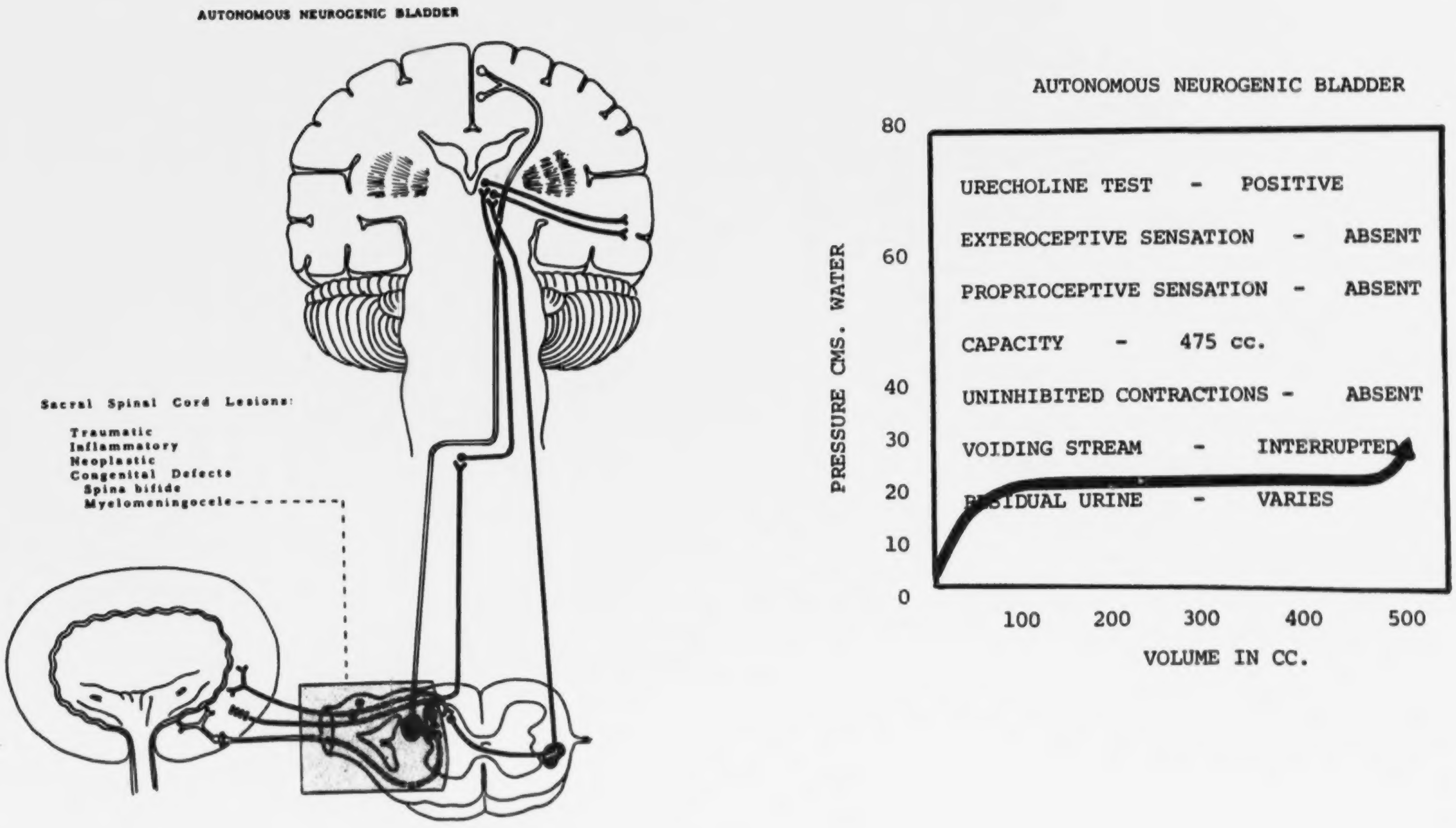

AUTONOMOUS NEUROGENIC BLADDER
Sacral Spinal Cord Lesions:
Traumatic
Inflammatory
Neoplastic
Congenital Defects
Spina bifide
Myelomeningocele
AUTONOMOUS NEUROGENIC BLADDER
80
60
40
30
20
10
0
PRESSURE CMS. WATER
100
200
300
400
500
VOLUME IN CC.
URECHOLINE TEST - POSITIVE
EXTEROCEPTIVE SENSATION - ABSENT
PROPRIOCEPTIVE SENSATION - ABSENT
CAPACITY - 475 cc.
UNINHIBITED CONTRACTIONS - ABSENT
VOIDING STREAM - INTERRUPTED
RESIDUAL URINE - VARIES

FIGURE 6.

The autonomous neurogenic bladder results from a lesion of all connections between the bladder and central nervous system. This type of bladder is seen when the second, third and fourth sacral segments have been destroyed. It is seen in spina bifida, meningomyelocele, trauma to the conus medullaris or cauda equina and in abdominal perineal resections of the rectum or other radical pelvic surgery. The patient has no sensation of voiding and is incontinent, usually of the overflow variety.

Cystometry reveals absent sensation. The bladder is usually hypotonic and without uninhibited contractions. Bladder capacity is normal or elevated and residual urine is usually elevated. The Urecholine test is positive. In addition, saddle sensation is impaired or absent and the bulbocavernous reflex is absent.

The patient with the autonomous neurogenic bladder, such as in congenital defects related to spina bifida and meningomyelocele can be managed by abdominal pressure. This method is satisfactory during infancy but emptying is not complete and intervesical pressure during this maneuver is very high. Eventually, the repeated increase of intervesical pressure leads to serious upper urinary tract disease and renal parenchymal destruction. Because of incontinence, upper urinary tract destruction caused by increased intervesical pressure and often superimposed infection, it becomes mandatory to divert the urinary stream in order to preserve renal function and make the patient socially acceptable. Unfortunately, the patient is frequently diverted too late, that is after chronic pyelonephritis and renal parenchymal destruction have occurred. Patients with spinal defects should be closely followed for evidence of upper urinary tract deterioration in order that urinary diversion may be carried out at the appropriate time.

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PROBLEMS OF FUNCTION

Harriet E. Gillette, M.D.

The entertaining, sociable child with glib phrases whose play periods are spent learning the intricacies of crutch walking will soon be the teenager who has passed the age during which definitive treatment is of benefit. Suddenly will come the awareness that school and play periods are finished, and the time has arrived for adoption of a constant pattern of living.

One measure of achievement of success is that per cent of capacity which is utilized. The intact person can find many excuses for his failure to employ all of his abilities. The individual with spina bifida has these and other valid reasons for his less-than-optimal performance. Some of these reasons stem from the same source as yours and mine; the majority are from real and measurable causes. It is the purpose of this presentation to delineate those causes as they affect the motor, intellectual, social and vocational spheres of the individual with spina bifida throughout his life span, and to indicate those factors which are amenable to treatment.

Problems to which the child with spina bifida is particularly subject are far-reaching, touching nearly all aspects of his life; unless corrected, or at least minimized, any one of them many be incapacitating. (Figure 1.)

	Motor	Intellectual	Social	Vocational
Incontinence	+		+	+
Parental Distress	+		+	+
Deprivation	+	+	+	+
Skin Breakdown	+	+	+	+
Impaired Intelligence		+	+	+
V/P Discrepancy		+	+	+
Perceptual Disturbances	+	+	+	+
Visuo-Motor Disturbances	+	+	+	+
Paralysis	+		+	+
Ataxia	+		+	+
Immobility	+		+	+
Fractures	+		+	+
Deformities	+		+	+

FIGURE 1

EFFECTS OF IMPAIRMENT

This is particularly true of incontinence of bladder and bowel; what ambulation is present may not be utilized because mobility may precipitate objectionable odors and appearances, or disturb collecting devices. Group activities, both in school and recreation are precluded, and employment away from home is impossible.

Complex emotional reactions of parents are initiated at first glimpse of the misshapen infant. All of the past of the parents becomes a part of their feeling toward their child, and in part determines whether or not they can love him. Resolution of their own problems may progress to the extremes of over-protection or rejection, one as devastating as the other, and as inimical to participation in sociality or gainful employment. Deprivation of many experiences normally encountered in the growing-up process, whether pleasurable or sad, is reflected in the difficulty in adapting to demands of a non-sheltering environment. Deprivation of sensory stimuli of all varieties, deprivation of stimulating intellectual exercises encourages that lump-like state designated "unmotivated."

The ever present threat of pressure sores determines participation in motor training programs, school and social activities, and regular attendance at employment. Decubiti are perhaps the costliest of the many complications in terms of prolonged inactivity, nursing care and frustration factor. While occasionally life-threatening, their chief importance lies in the frequent and prolonged interruptions of progress.

Discrepancy in verbal and performance scores on psychologic examination contributes to the lack of understanding of the child by those around him, for the irregularities of mental functioning limit the areas in which mental abilities may be utilized. Disorders of perception are particularly troublesome, influencing motor and intellectual performance. A disorder of body image concept and lack of orientation to space results in an appropriate caution regarding position changes, which may be interpreted by others as fear, apathy or stupidity. It is difficult to understand why a child fails in efforts to learn to use crutches until it is found that his body schema is so constructed that his legs have minimal representation, and he is unable to perceive an extension of force from arms through crutches to the floor for support for the body. Lack of visuo-motor coordination produces difficulty in manipulation and construction. The importance of perceptual disability cannot be overemphasized, for its impact can be seen in functioning in all aspects of performance.

Those problems of function which deal chiefly with motor performance are the most obvious, yet probably of the least significance in total performance. Paralysis which is minor in degree may limit motor performance only to the extent that the child finds himself in the never-never-land of the non-handicapped and the non-achiever. Sparing of certain neuro-motor units produces imbalance of power which is the forerunner of skeletal deformity. Lack of stress upon bones with subsequent demineralization, deficient sensation and unusual stresses combine to allow ease of fracture, so that a cast on one or another leg may be accepted as a way of life. Interference with motor performance may be solely a result of central impairment of equilibrium responses; there may be the added factor of perceptual disability. Either may negate the effectiveness of good motor control.

ALLEVIATION OF DISABILITIES

The imposing list of impairments compels consideration of some disturbing points, relevant to the problem of treatment.

Prior to the present era of a concerted effort to reduce disability in spina bifida, did many children survive, and did any become self-sufficient? It must be acknowledged that custodial institutions can testify to the survival rate, for it has been customary to remand such a child to the state training school without delay--or, it may be said, without due consideration. It seems quite remarkable, in the light of numerous associated handicaps, that a goodly number of adults with spina bifida are entirely self-sustaining. Their parents deserve acclaim for their understanding, patience and ingenuity. It is because of the potential for a productive, meaningful life that an effort is being made to minimize the handicap whenever possible, through application of sound treatment principles.

The major impairments to optimal performance have been enumerated, and their far-reaching consequences briefly described. How many of these can be alleviated? (Figure 2.)

	Motor	Intellectual	Social	Vocational
Incontinence	(+)		(+)	(+)
Parental Distress	(+)		(+)	(+)
Deprivation	(+)	(+)	(+)	(+)
Skin Breakdown	(+)		(+)	(+)
Impaired Intelligence		+	+	+
V/P Discrepancy		+	+	++
Perceptual Disturbances	+ ?	+ ?	+ ?	+ ?
Visuo-Motor Disturbances	+ ?	+ ?	+ ?	+ ?
Paralysis	+ ?	+ ?	+ ?	+ ?
Ataxia	+		+	+
Immobility	(+)		(+)	(+)
Fractures	(+)		(+)	(+)
Deformities	(+)		(+)	(+)

FIGURE 2

IMPAIRMENTS AMENABLE TO TREATMENT

Incontinence of bladder and bowel need not limit function if techniques described by our colleagues in urology are applied; morbidity and mortality due to renal disease should be lessened with the proper and prompt surveillance of the urinary system .

Distress of parents is a normal reaction, but with support, encouragement and above all with the offer of help for the child, this sadness can be kept within bounds, and mental illness prevented. What may appear to be an expensive program of treatment procedures during the few early years of the child's life is very little compared to the long-term care of a decompensated parent, with the many social ills attendant upon a disorganized household.

The team effort is essential for prevention of deprivation in motor, intellectual, social and vocational spheres. From the first few days of life, when the infant is subject to imprinting of the mother figure, through the stage of development of a mechanism needed for movement, through the intellectually demanding school years and on into appropriate vocational placement, each member of the treatment team plays a vital role in salvaging the diverse abilities of the child. Deprivation in any one of the spheres will minimize performance in all the others, and no effort is too great in exercising prevention.

Skin breakdown is an indication of either ignorance or neglect, and cannot be tolerated. Since there is only one causative factor, that of pressure on devitalized skin, there is only one principle of prevention; the rule of no prolonged localized pressure points over bony prominences may be invoked at will.

On this day, can it be said with assurance that proper tools are at hand for accurate assessment of that entity termed intelligence? Can that nebulous complex be defined, or its separate parts identified? Does interpretation of sensory input, commonly called perception, have a significant correlation with reasoning ability? Can perception be equated with performance of skilled and unskilled motor acts?

That level of intellectual functioning which results from the best efforts to salvage all possible neural elements is the major determinant of appropriate social and vocational placement. Adjustment to that placement is variable, and is dependent upon the expertise of all those who are in contact with the individual and his milieu. In this respect, the employment of a given intellectual level is a variable, and amenable to treatment, even though intelligence per se may not be.

Training in ambulation and in performance of self-care activities may be difficult, prolonged and sometimes unsuccessful. Frequently the therapist reports that there is excessive fear of falling, or that there is difficulty in finding the foot on which to pull a sock. If such a child is asked to draw a picture of himself, the resulting figure is quite apt to appear with shortened or misplaced legs; small wonder that he is unable to use a body part which is inadequately represented in the body schema. Tests of spatial orientation yield results indicating disorganized responses in this area; perhaps the child's apprehension of moving about is appropriate caution rather than malignant fear or ataxia. Continuing investigation of perception may produce more definitive methods of testing and of training; if so, performance in all spheres will be amenable to treatment.

Mechanical factors concerned with lack of muscle power or with unbalanced muscle action, contractures and their resultant skeletal deformities can and should be favorably influenced through preventive measures. Lack of motor units can be supplemented by orthotic devices; their employment in functional activities, however, is dependent upon a voluntary effort and optimal cortical functioning. Orthopedic surgery plays an important role in providing the necessary alignment of the locomotor apparatus so that neuro-muscular units can act in a purposeful manner. This aspect of care must be continuing throughout the growth period; it is essential that it be correlated with other aspects of care. (Figure 3)

	Motor	Intellectual	Social	Vocational
Incontinence				
Parental Distress				
Deprivation				
Skin Breakdown				
Impaired Intelligence		+	+	+
V/P Discrepancy		+	+	+
Perceptual Disturbances	?	?	?	?
Visuo-Motor Disturbances	?	?	?	?
Paralysis				
Ataxia				
Immobility				
Fractures				
Deformities				

FIGURE

REMAINING FIXED IMPAIRMENTS

The majority of the problems of function are amenable to treatment; those which at the present time appear to be fixed or of questionable accessibility to treatment, lie in the intellectual sphere. Even those can be handled in such a manner that their significance is not magnified, or their degree increased.

DEVELOPMENTAL DISABILITIES AND PROBLEMS OF INTELLECT*

John H, Meier, Ph.D.

The human brain is a many splendored thing. Where else can you find a computer which has several billion flip-flop circuits, occupies less than one cubic foot of space, operates on the energy of a peanut for up to four hours, is completely mobile, and is produced with unskilled labor? Regardless of the response to this rhetorical question one cannot dismiss the enormous complexity of the human brain which performs the very intricate functions of intellect. Many might argue that the intellectual functioning of the human organism is far more elegant than that which is simulated by a computer; others might argue that the whole realm of affect is ignored when such a crippled cognitive analogy is drawn; and still others have taken serious issue, on the basis of personal competence and/or theological convictions, with the notion of the unskilled labor involved in producing such a marvelous mechanism.

Related to the fundamental questions of the origin and nature of human intellect are the intensely controversal considerations involved in the measurement of intellectual functioning. Intelligence is a term which refers to the measured state of a person's intellectual functioning at any given time and has been somewhat facetiously, albeit operationally, defined as that which intelligence tests measure. The very foundation of psychometry rests on the somewhat shifting sands of the physical sciences which typically operate on the assumption that insofar as anything exists it can be measured; even such an ephermeral phenomenon as love is fathomed by some scientific romanticists to be as deep as the ocean, etc. An intelligence quotient (IQ) is a single numerical index which quantitatively signifies how knowledgeable a person is relative to his chronological age and may be used to compare his intelligence with that of the majority (norms) of other individuals. Because of its relative statility over time (Bloom, 1964), the IQ, in turn, is frequently used for sorting individuals into classifications of relative ability to profit from educational experiences which are similar to those which intelligence tests measure. Of course the predictive power of IQ in terms of school success is probably due at least as much to the relative stability of the environment as it is due to the alleged immutability of brain function (Wechsler, 1966).

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*Paper presented at a Symposium on Spina Bifida held at the University of Colorado Medical Center, November 24-26, 1969. The author is director of the John F. Kennedy Child Development Center and Associate Professor of Pediatrics and Clinical Psychology at UCMC.

The above somewhat circular exposition reveals a problem with the concept of intellect in addition to problems of intellectual functioning manifest in individuals with developmental disabilities such as spina bifida and such related conditions as meningomyelocele and hydrocephalus. It points up a crucial qualitative consideration which is seldom elaborated. In short, intellect is not a unitary quantity any more than is CNS functioning, which demands thorough differential analysis for complete understanding (Meier & Martin, 1969). Guilford (1959) has proposed a model of intellect, synthesized from compelling empirical findings, which has at least one hundred identifiable factors, many of which are not sampled in traditional intelligence testing. The age-old controversy about the relative contribution of heredity and environment to the stability and magnitude of intelligence is a perennial controversy highlighted most recently by the claims of Jensen (1969) and the many counter-claims these have precipitated. Although the scope of this paper does not permit further discussion of the aforementioned problems with the concept of intellect itself, it is necessary that any simplistic notions be dispelled at the outset.

Since there is a fairly high relationship between the incidence and repair of spina bifida and related meningomyelocele with hydrocephalic conditions, which in turn are characterized by various intellectual deficits, two models of communication systems are presented in order to depict some of the interrelationships. Although these networks are nearly self-explanatory, it is exigent that several of their salient features be briefly described and related to the problems of intellect associated with these conditions. Figure 1 depicts a communications network in which such a paper as this one fits. This Symposium on Spina Bifida is held to effect accurate and current interdisciplinary communication among the numerous and scattered professionals who are dealing with the condition.

The transmitters (numbered 3 in Figure 1) share their recent research and clinical findings as well as theoretical constructs (1) with fellow workers (3,7, and 8) in the field. In any communication system there are various filters which condition the information by eliminating what might be referred to as noise (2 and other wavy intersecting lines) in the system. Noise to one reporter or listener may be music to another; the transmitter, who may be a neurosurgeon or an anthropologist, is tuned to certain phenomena and records and reports data from his biased point of view. The remainder of the network indicates additional points of possible distortion which typically bias the communication of facts and ideas among and across the several disciplines concerned with the issues at hand. A symposium such as this serves to short-circuit the network by enabling direct dialogue between, for example, practicing physical therapists (8) and a research physiatrist (3). Such direct interchange and opportunities for clarification help bridge some of the gaps in communication which frequently arise from technical/statistical language, or esoteric semantic fuzziness, or inavailability of appropriate materials, or inadequate sampling of the available resources, or an inability to translate research theory into clinical practice.

Figure 1. AN INTER-INDIVIDUAL COMMUNICATIONS NETWORK

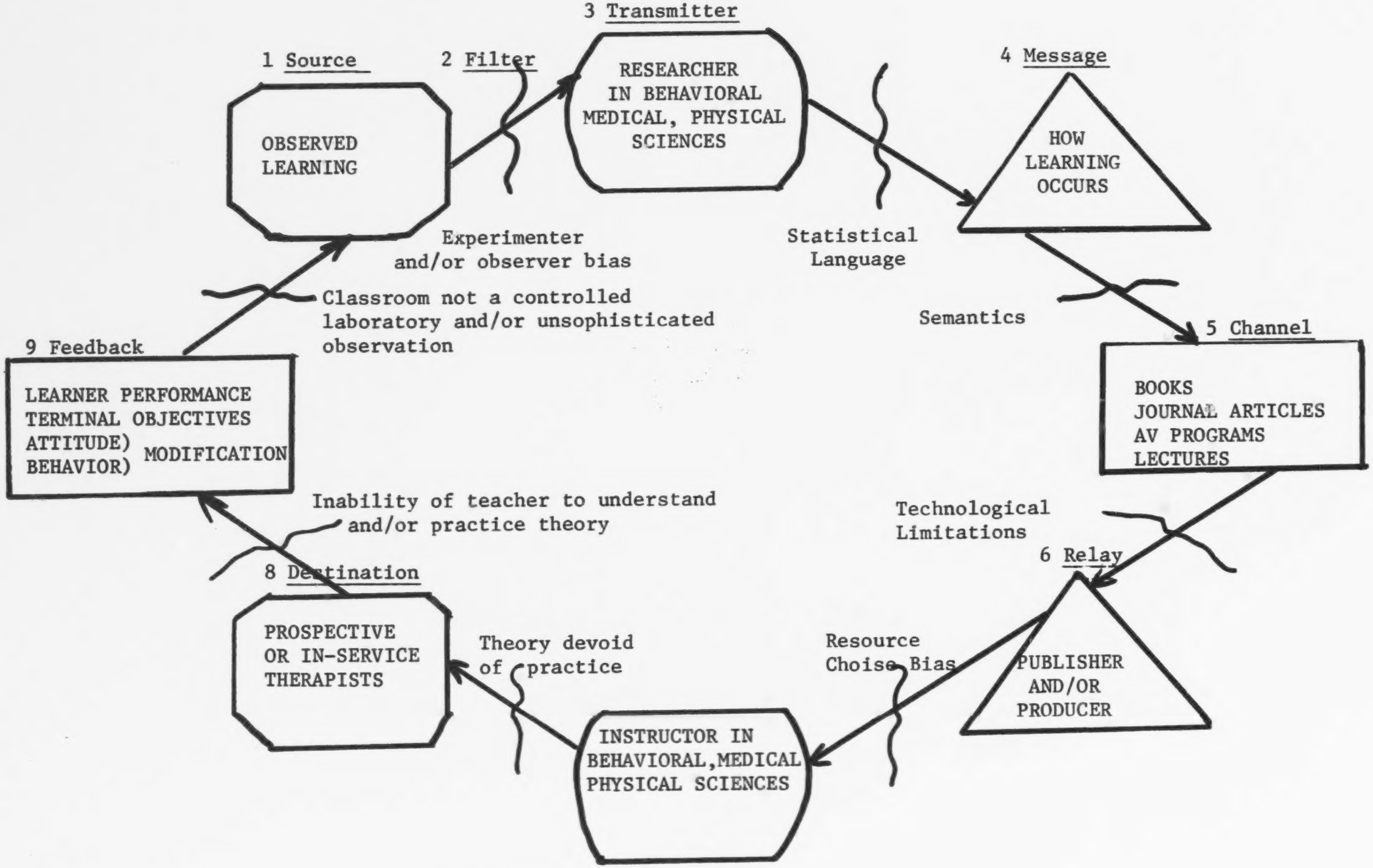

1 Source
OBSERVED LEARNING
2 Filter
3 Transmitter
RESEARCHER IN BEHAVIORAL MEDICAL, PHYSICAL SCIENCES
4 Message
HOW LEARNING OCCURS
Statistical Language
Semantics
5 Channel
BOOKS
JOURNAL ARTICLES
AV PROGRAMS
LECTURES
Technological Limitations
6 Relay
PUBLISHER AND/OR PRODUCER
Resource Choise Bias
INSTRUCTOR IN BEHAVIORAL, MEDICAL PHYSICAL SCIENCES
Theory devoid of practice
8 Destination
PROSPECTIVE OR IN-SERVICE THERAPISTS
Inability of teacher to understand and/or practice theory
9 Feedback
LEARNER PERFORMANCE
TERMINAL OBJECTIVES
ATTITUDE) MODIFICATION
BEHAVIOR)
Classroom not a controlled laboratory and/or unsophisticated observation
Experimenter and/or observer bias

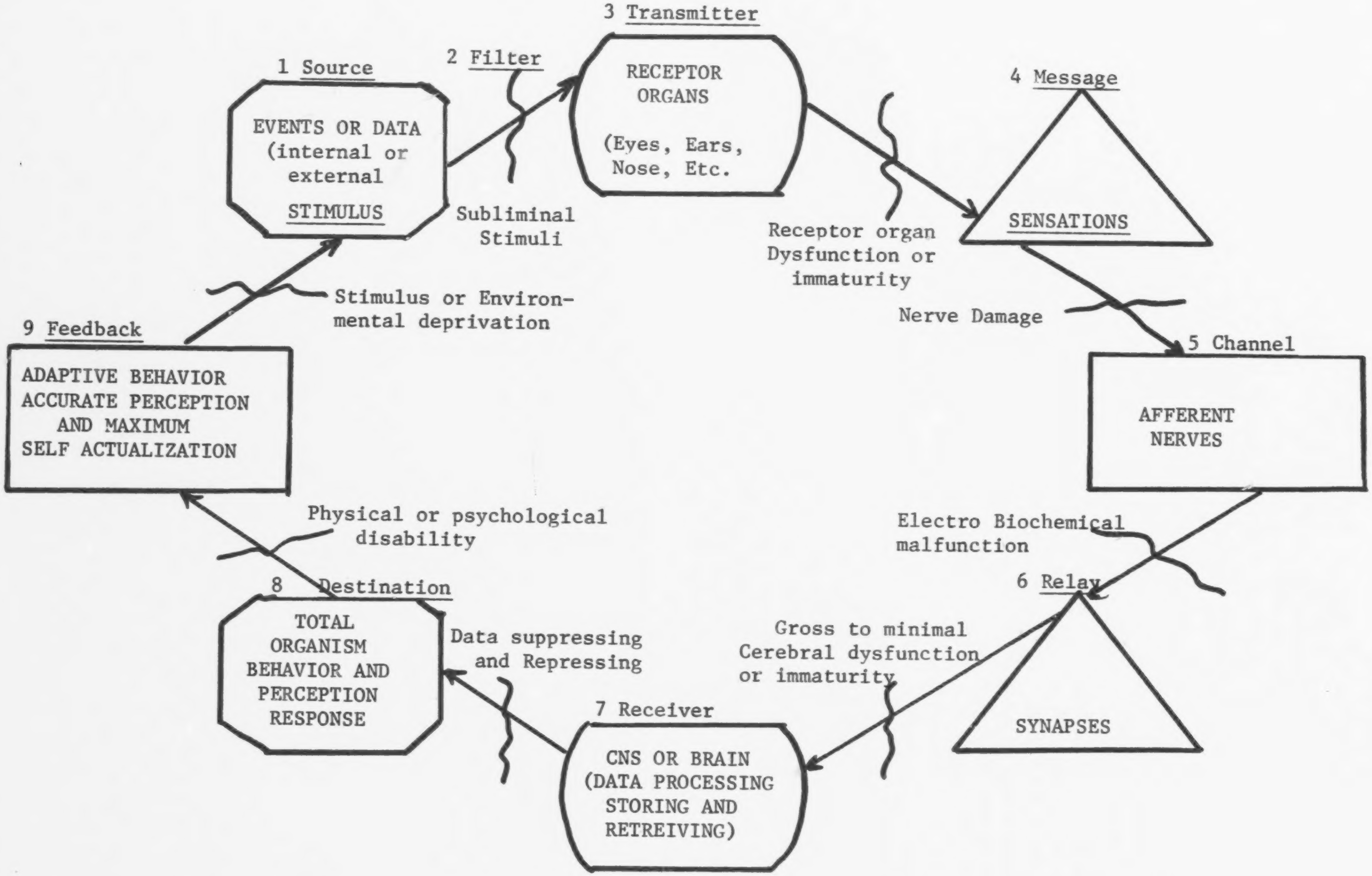
Figure 2 depicts a single human organism as a communications network with the same basic system components. As an illustration of some of the problems of intellect related to spina bifida, a child with an hydrocephalic condition is herein considered. Moreover, since one of the primary manifestations of intellectual deficit in hydrocephalic children is that of communication disorders, a child with an auditory communication disorder is plugged into this communications system to serve as an example.

As various stimuli (1) from the internal and/or external environment of the individual impinge on the receptor organs (3, the ears) and, if these stimuli are of sufficient intensity, that is above the threshold required to trigger the receptor nerves, a message or series of sensations (4) is generated. Subliminal stimuli (2) are technically those which in fact exist in the environment but are too weak to trigger the receptors; non-technically, subliminal advertising uses stimuli too fleeting to be consciously available but does seem to have some unconscious and often controversial impact. The reception of such stimuli is also dependent upon the relative acuity of the receptor organs; an inaudible (to humans) sound is not sensed because of the limitation of the human hearing apparatus itself, whereas a normally audible sound may not be heard because of a dysfunction of the hearing apparatus. There are many audible sounds which are sensed but in fact are not consciously or unconsciously acted upon because of the ability of the other portions of the communication system to selectively attend to only certain sound patterns and exclude or turn off other irrelevant or meaningless sounds. It has been demonstrated that a certain optimal level of distracting sounds in the background forces the organism to attend even more closely to the salient stimuli in the environment. A person may hear better when there is an optimal level of competing stimuli which he must selectively eliminate and eventually habituate; for example, limited static on a radio program (snow on a TV program) may cause one to attend more carefully to sort out the message. The manner in which and the extent to which the receptor organ is able to discriminate is not fully understood. Evidence is now mounting that the retina, and perhaps other peripheral transmitters, such as the cochlea, in addition to converting data into neural impluses, does some preliminary processing of the data before sending the message on through the network.

After a message (4) is generated it must flow smoothly through the peripheral (5) into the central (6) nervous system. Provided that there is no nerve damage (breakdown at the cochlear analytic level thus eliminating high tones, myelitis, etc.) or electro-biochemical malfunction (faulty synaptic transmission; RNA, ascorbic or folic acid deficiencies; nuclei in the brain stem damaged by deposits of blood cell pigment in a Rh athetoid condition; reticular activating system disorder, etc.),the message is received by the brain (7).

It is noteworthy that auditory dysfunction due to any number or combination of these conditions is typically less likely to be noticed than analogous problems in the visual communications system. This is probably because visual problems are more obvious than are auditory ones. Conditions such as otitis media have been found in many children who were singled out as having difficulty learning for unknown reasons (Meier, 1969). Such children, as well as their peers and adults, frequently are unaware of the fact that

Figure 2. -AN INTRA-INDIVIDUAL COMMUNICATIONS NETWORK
DEVELOPMENTAL DISABILITIES AND PROBLEMS OF INTELLECT


1 Source
EVENTS OR DATA (internal or external
STIMULUS
2 Filter
Subliminal Stimuli
3 Transmitter
RECEPTOR ORGANS
(Eyes, Ears, Nose, Etc.
Receptor organ Dysfunction or immaturity
4 Message
SENSATIONS
Nerve Damage
5 Channel
AFFERENT NERVES
Electro Biochemical malfunction
6 Relay
SYNAPSES
Gross to minimal Cerebral dysfunction or immaturity
7 Receiver
CNS OR BRAIN (DATA PROCESSING STORING AND RETREIVING)
Data suppressing and Repressing
8 Destination
TOTAL ORGANISM BEHAVIOR AND PERCEPTION RESPONSE
Physical or psychological disability
9 Feedback
ADAPTIVE BEHAVIOR ACCURATE PERCEPTION AND MAXIMUM SELF ACTUALIZATION
Stimulus or Environmental deprivation

they are not hearing normally and may find the results of this diminished acuity to be most frustrating when they fail to follow directions or perform other tasks which are verbally conveyed to them, their siblings, and classmates. Thus, a secondary result of a specific auditory disorder is often a mild distrust of everything one hears to a severe emotional emotional disturbance arising from apparently irrational reprimands for not properly executing oral requests which were improperly perceived auditorially.

Difficulties in the reticular activating system may prevent the child from being able to selectively attend to certain desired auditory stimuli since all stimuli seem to have the same significance to him. The paradoxical affect of certain amphetamines on children with disorders described as distractability and hyperkinesis may in fact enable these children to better sort out the stimuli to which they wish to attend, thus improving their concentration on the task and reducing their distractability and hyper-activity, which may represent random and sometimes frantic scanning of the auditory (visual, etc.) field for significant events.

However, the cortical areas of the brain which are primarily responsible for auditory processes may be damaged or developing slowly, thus making the differentiation or discrimination of various sound patterns quite poor or ambiguous; such fuzzy reception (decoding) of data necessarily renders the associating and expression (encoding) of the same date equally fuzzy. There is an acronym used in computer technology referred to as GIGO, which stands for Garbage In Garbage Out. The GIGO phenomenon refers to the fact that garbled reception of auditory stimuli inevitably results in improper or defective associations and expression of the original input or new combinations thereof.

The flood of incoming data - billions of bits per minute - is initially processed in terms of basic survival (involuntary) needs and drives. The individual's cognitive and affective domain play a major role in determining which messages are worthy of conscious attention and which are to be ignored, suppressed, and even repressed. There is some evidence that much of the data received by the organism are stored and available for later retrieval even though conscious reflection sometimes does not yield the memory of certain experiences. Before removing certain diseased portions of the brain, a neurosurgeon maps the intact areas and can evoke memories of a musical selection, for example, when an electrical impulse is applied to a specific needle point area in the conscious brain.

From a psychoanalytic viewpoint (with some tentative neuro-physiological considerations mentioned parenthetically) the pleasure-seeking, pain-avoiding organism filters out those messages which emanate from or are in harmony with the libidinous instincts (located in the old brain or rhinencephalon) and they are not allowed to enter into consciousness by the executive ego (hypothalamic structures or mesencephalon), because of the super ego's (cortical structures) frame of reference which prevents certain messages from being related into consciousness or from being reflected upon if they seep through.

From a cognitive-field point of view, some messages cause cognitive dissonance (disequilibrium and increased tension in the life space) to the extent referred to as information overload in the computer system; in such cases, the receiver simply turns off or awaits, perhaps seeks, a more compatible (less dissonant) or more meaningful message which doesn't do such violence to the status quo, doesn't require unbearable amounts of restructuring (new learning) in the life space and doesn't frustrate the learning process with meaningless noise.

Regardless of the course of data through the learning organism, which has been dismissed as a black box by many investigators, a relationship can be established between a specific input (1) and an observable output (8) which is either appropriate or inappropriate in terms of normal behavior (9). Intelligence testing typically introduces a controlled stimulus (1) and measures the corresponding response (8). When the response to a standard stimulus is quite deviant from the average, some exception is noted and a disorder in the system is inferred. If a person is in a real life situation where danger is signified by an auditory warning, such as a siren or a rapidly approaching ambulance, and the person makes no effort to avoid the impending harm, he is neither orthopedically handicapped nor suicidal, it is judged that some hearing or auditory disorder is present.

When this is the case, a differential diagnosis is indicated to determine where in the communications network a malfunction is occurring. Such a diagnosis may begin with the patient's history to determine whether or not the auditory system is underdeveloped as a function of the environment, since stimulus or environmental deprivation is a factor contributing to inadequate visual and auditory development (Deutsch, 1967). Valverde (1968) and others have clearly demonstrated that environmental or stimulus deprivation can reduce the number of dendritic spines in the visual cortex of experimental animals and one could legitimately assume that the same phenomenon would be found in the auditory cortex of animals and humans. Environmental enrichment, on the other hand, has a reversed effect on visual cortex development (Rosensweig, 1966).

Since hydrocephalic children frequently have defective verbal behavior, it is conceivable that the difficulties have their etiology to some extent in the general deprivation many of these children suffer simply due to the more limited auditory and visual experiences they have, particularly early in life, due to restricted verbal and social interaction as a result of their handicap (Myklebust, 1965). One of the most significant differentiations in their verbal disabilities is that, although they are typically quite talkative and fluent, they seem unable to make appropriate verbal responses to anything other than the most superficial conversational statements.

> Commonly found were observations that these children were unable to stay on a specific topic, that they tended to engage in social conversation rather than use language in a meaningful or informational manner, and that they would relate a topic to their own personal experience and get carried away by these personal associations. (Fleming, 1968, p. 75-76).

Appropriateness of Response to the Children's Apperception Test pictures was a category in which the hydrocephalic group of children with near normal and normal intelligence quotients had a median percentage of 14% inappropriate responses compared to the matched control group median of 5% which was a significant difference (.05). This is consistent with the suggestion by Laurence and Coates (1962) that in many types of brain damage in children the vocabulary is near normal in contrast to an impaired reasoning ability. One of the most common inappropriate types of remarks included various conversational observations about the testing environment or the investigator and many statements totally unrelated to the situation or the task. It is suggested by Fleming (1968) that:

> ... These children were not so much distractable in the passive sense, but would actively seek out other topics from the environmental surroundings;

he further observes that:

> many of these children appeared to seek distraction from the required task of picture description but would attempt to maintain the flow of language and the conversational relationship. (p.80)

Several other workers have described the motor performance and verbal and social behavior of hydrocephalic children.

> Patients with hydrocephalus exhibit a distinct pattern of neurological signs, with spasticity of the legs and, to a lesser extent, of the arms as the most constant feature, and with ataxia, tremor, imbalance, and clumsiness of fine finger movements usually also present. This has been noted previously and Ingram and Naunton (1962) regarded the motor dysfunction as a combination of a cerebellar ataxia and a spastic diplegia. Apart from the lowering of IQ, the intellectual effect of hydrocephalus seems to be less constant. The characteristic most frequently shown is the striking development of an "auditory menory" in otherwise retarded children (Ford, 1960), and the shallow intellect often associated with talkativeness described as the "chatterbox syndrome" by Ingram and Naunton (1962) and the "Cocktail party syndrome" by Hadenius and her co-workers (1962). Hagberg and Sjogren (1966) regard these features, both motor and intellectual, of hydrocephalic patients to be so characteristic that they have designated them as "The chronic brain syndrome of infantile hydrocephalus" (1962). In some hydrocephalics where there is no obvious intellectual impairment detected by methods at our disposal, nor any motor abnormality, more subtle changes show themselves as emotional lability or lack of self reliance. The more severe the hydrocephalus, both in this series and in that of Hagberg and his collaborators (1966), the more marked were the motor disabilities, the mental retardation, but to a much lesser extent the intellectual characteristics. (Laurence, 1969, p.80)

> Hydrocephalic children were often found to be mentally retarded but educable, with a peculiar contrast between a good ability to learn words and talk and not knowing what they were talking about. They loved to chatter but think illogically...(Hadenius, et al., 1962, p. 118).

> They make a certain amount of headway with motor skills, with abilities to talk, sing,memorize, and be "cute," but then a discouragingly high percentage of such youngsters gradually stop advancing (Matson, 1961, P. 450).

> These observations, coupled with the findings of this study, might well indicate that some hydrocephalic children appear to be bright on the basis of their verbal and social behavior, but that frequently academic performance and formal testing fail to support this impression. Unfortunately, what is "cute" or appropriate behavior at one age becomes a social impediment later on in life when more mature behavior is expected. Unrealistic expectations may also lead to continued failure and disappointment in academic work unless any discrepancy between apparent verbal social abilities and those involved in more meaningful and appropriate language behavior is realized (Fleming, 1968, p.80).

While bemoaning the dearth of good statistics regarding spina bifida, Tizard (1968, p.1) cites several studies reporting children who survive until school age and describes some of their performance in both the social and educational realms. Of the estimated two per one thousand births which are characterized by spina bifida, Tizard suggests that the majority of such children who reach school age are educable in terms of IQ and have responsed to education. He cites a study by Stephen (1963) in which twelve out of twenty-five spina bifida children were making above average progress, twelve were making average progress, and one was making below average progress in a school for the physically- handicapped. Of seventy who had attended one or other of two schools, fifty had IQ's of 80 or above. Beks, et al, present the results of neurosurgical treatment in 133 patients born with spina bifida.

> Some 57% developed hydrocephalus following closure of the cele. Of the 98 surviving children, 95 were available for an exhaustive follow-up, which showed that 62 (65%) had no or only slight disturbances in walking.

> The intellectual development of children treated for spina bifida is practically normal, provided the hydrocephalus which may develop afterwards can be treated early and successfully. There are also indications that the prognosis of hydrocephalus which develops after closure of the cele, is more favorable in terms of cerebral functions than that of a comparable degree of hydrocephalus resulting from other causes. In the latter group, therefore, another noxious factor must play a role besides the hydrocephalus.

> We believe that children suffering from spina bifida must be treated. In our opinion, a strong argument in favor of this view is the potentially normal intelligence, which is hardly affected by the presence of a mild form of hydrocephalus. (1969, P. 416)

Eckstein and MacNab (1966) reported that in their series of studies of fifty-six children with meningomyelocele and about whom information was available and who were of school age, twenty-five, or 45%, were in normal schools and twenty-six in schools for physically-handicapped children. Only three were in institutions, one was uneducable, and one was receiving home tuition.

> These findings indicate that substantial numbers of educable children with meningomyelocele survive to school age and adolescence and that the task of educating them is one that can be successfully tackled. The problems of schooling and the difficulties of adjustment in adolescence and adult life are likely to be very serious -- but they are of a kind that are faced by other handicapped persons, and the great need is to provide adequate services (Tizard, 1968, p. 3).

Among some of the services required, Tizard mentions family counseling to enable parents of children with spina bifida to cope with the problems of management and the worry and uncertainty about the child's future, particularly during the child's infancy. He also mentions:

> A child who is handicapped in any way is likely to have some difficulties in passing his milestones, not only because of the direct consequences of his handicap, but also because he does not have ordinary opportunities to explore his environment, to enjoy the normal range of experience, to play with other children and his parents.... Where the child has multiple handicaps the task of the educator and the parents is to provide as wide a range of compensatory experience as possible (1968, pp. 3-4).

These studies and many others investigating similar phenomena tend to point out the nature of the handicap of intellect which frequently follows spina bifida and its associated developmental disabilities. The notion that only the abstract reasoning and problem-solving portions of intellect are impaired or destroyed in many cases is encouraging in terms of the prognosis for the patient to get along in the everyday concrete kinds of language exchange necessary for survival. However, it indicates the potentially misleading characteristic of an apparently normally adjusted individual who, in fact, has real difficulty relating abstract ideas and acting appropriately upon their consequences. An attractive hypothesis is that the sequelae of spina bifida in terms of intellectual functioning, particularly in terms of the hydrocephalus discussed herein, may be that the individual is more or less restricted to the concrete level of language operations which would be regarded as a less mature stage of development by investigators such as Piaget (1952). Language is one of the last, and certainly most distinguishing characteristics to develop in the evolution of species. It is conceivable that abstract intellectual functioning, being theoretically more advanced than concrete language operations, may also be the first to be rendered defective due to environmental deficiencies or physical trauma and maldevelopment. It may be the most delicate or vulnerable human characteristic and thus the first to fail under adverse conditions such as increased intercranial pressure, reduced circulation, blows to the head with a very thin cerebral mantle, etc. Since tests of intelligence don't sample higher-order cortical functioning until the child is well into school, the apparent attenuation of intellectual progress is largely a result of the changing characteristics of intellectual progress is largely a result of the changing characteristics of intelligence sampled in older people. Although rote memory such as that tested by vocabulary or repeating digits is still an important fundamental skill, the ability to reason and solve problems becomes increasingly salient particularly for academic achievement.(Meier, 1965). It must also be borne in mind that the elegance of subtest patterns is lost in a single IQ score. The scatter of subtest scores may vary greatly within the same individual's life span as verbal subtests, for example, begin to sample increasingly abstract abilities for older individuals and cease to rely so much on rather automatic language (McCarthy and Kirk, 1968). Whereas vocabulary and rote information offset low performance in other areas (sensory-motor-perceptual) while the child was young, their diminished value later cannot compensate for plateauing or non-development of higher order synthesizing and analyzing skills.

In addition to the observation that such developmentally handicapped children are frequently prevented from vigorous and meaningful interaction with their environment by their parents or other responsible adults, the possibility of Rosenthal's (1968) prophecy-fulfilling phenomenon should also be considered insofar as it could act to cause adults who are responsible for the education of such children to expect considerably less of them than their real intellectual potential warrants and to act correspondingly more protective and less stimulating with regard to their intellectual development. Perhaps part of the reason for these children failing to develop abstract problem-solving abilities is that they are not expected to nor taught how -- an intriguing hypothesis which warrants further investigation, in spite of the finding that, "The neuropathologic examination of 359 brains from patients at a hospital for the mentally retarded revealed that 83% of them had moephologic disorders accounting for the clinical deficits." (Freytag and Lindenberg, 1967, p. 391). Lorber (1968) concludes a report on the early treatment of extreme hydrocephalus with the following statement:

> These results suggest that no case with primary congenital hydrocephalus should be considered hopeless and all should have the benefit of the best treatment, irrespective of the degree of the hydrocephalus, because the majority will do well if treated in the first few months of life. Even the most extreme degrees of hydrocephalus are compatible with normal physical development, a normal sized head and superior intelligence if operative treatment is not delayed. (p.27.)

Foltz (1968) also reports a very optimistic outlook for treated hydrocephalics:

> Comparison of 56 children in our study group with the 81 in their group is striking. Fifty-one percent of the treated hydrocephalics had a normal IQ range of 90 to 110 whereas only 23% of the untreated hydrocephalics had such an IQ. This statistically significant study strongly implies that the normal intellectual capacity of the hydrocephalic brain can be attained if the hydrocephalus is controlled. Uncontrolled hydrocephalus in a child favors less intellectual capacity. (p. 450).

As Lorber points out in his article, the added complication of meningomyelocele accompanying spina bifida makes the prognosis somewhat less optimistic but not hopeless. In another article dealing with the assessment of school placement in children suffering from encephalocele and meningomyelocele, the following corroborating statements were made:

> An assessment was made of the physical handicap and intellectual capacity of seventy children with meningomyeloecle and encephalocele born in Liverpool in the years 1960 and 1963 in order to ascertain the future educational needs of such children. A third of the children were considered fit to attend ordinary school. Approximately half of the children required educating in a school for physically-handicapped pupils. The remaining children will need other special provisions on account of associated handicap such as blindness or severe subnormality. (Burns, 1967, p. 29)

To give some notion of the range of intellectual capacity of these seventy children, it varied from a developmental quotient of less than 20 to a Stanford-Binet IQ of 148, the average IQ being 87 for the sixty-four children who were considered suitable for school. However, it is noteworthy that these children ranged from 4 to 6 years of age, and the preceding cautions are germane as they grow older and might be expected to perform more abstract intellectual functions. Some of the defects of the special senses, as depicted in the communications network (Figure 2, Number 3), included several visual defects such as total blindness, strabismus, and astigmatism. Speech defects (9) included babbling and single-word speech, and developmental dyslalia. Of the sixty-four children with meningomyelocele or encephalocele deemed educable, there was no evidence of hearing defects and only one case of active otitis media. A brief summary of the school placement indicates that six of the children were so grossly handicapped as to be unsuitable for school,twenty-three were capable of attending ordinary school,thirty-eight with multiple disabilities were placed in schools for the physically handicapped, one was placed in a school for educationally subnormal children, and two in a school for the blind.

> It is interesting to note the relationship between intellectual handicap, physical disability, and school placement. Thirty-nine children had intelligence quotients between 50 and 89, but only eight will be physically fit to attent normal school at five. Of twenty-five children with IQs from 90 to 120 and above, fifteen are physically fit to attend normal school and nine of these have not been readmitted to hospitals since their primary closure. (Burns, 1967, p.27)

The same author then discusses some of the educational implications of such physical handicaps. He makes some specific recommendations for Liverpool and then adds several points:

> The school will admit all types of physically-handicapped children, but predominantly these would be children with meningomyeloceles....The school will include provision for a nursery unit of 20 children from the ages of three to five years. Preschool nursery training is of inestimable value to physically-handicapped children.
>
> The educational requirements of these children are similar to children attending ordinary school of comparable age and intelligence, but their medical requirements mean that ancillary staff able to cope with incontinence, physiotherapists to encourage children in walking, nursing staff to deal with pressure sores, etc., are essential.... The aim of the school should be to make full use of the child's intellectual potential, to encourage the child to become as mobile as possible, and learn to accommodate to the handicap. The school also has a supportive role in helping the parents in the management of these severely handicapped children. (Burns, 1967, p. 28-29)

The writer strongly concurs with the preceding statements regarding educational programming for handicapped children. The value placed upon preschool education is particularly noteworthy. Several attempts to intervene with compensatory educational programs for infants and young developmentally retarded children have yielded very promising results (Meier, 1967; 1968; Nimnicht, McAfee, and Meier, 1969; Meier, 1970a; and 1970b).

This completes the full circle in the communications network, and the preceding representative but by no means exhaustive sample of the current theoretical and practical state of the art makes it clear that the problems of intellect manifest in children with spina bifida are quite complex, especially when it is accompanied by the common complications of meningomyelocele and hydrocephalus. The behavioral, medical, and physical sciences are consequently called upon, not only to provide earlier identification and more thorough interdisciplinary evaluations of these conditions in their incipiency, but also to plan realistic remedial programs which will enable the individual to realize his maximum potential. The treatment may take whatever form is necessary to eliminate or compensate for the various sources of noise in the communications network which act as deterrents to the organism's getting, processing, and acting upon complete and accurate data. It is axiomatic that, the more that is done to eliminate noise in the two systems of communication among professionals and within individuals and the sooner that it is accomplished, the better is the prognosis for those handicapped by lack of professional information or by birth defects caused by any one or combination of these conditions and their attendant subpar professional practice or a child's developmental disabilities and problems of intellect.

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SOCIO-ECONOMIC PROBLEMS

Howard G. Rosen, A.C.S.W.

Thanks to the progress of science, man's average life span is longer than in previous eras. This is also true of children born with physical and/or mental handicaps. It means that a greater number of these children will survive and form part of our school age and adult society, thus providing us with challenges.

Until fairly recently the fate of handicapped children was harsh if not cruel, and only gradually are we beginning to realize that they must be assured of a fair chance of a satisfying life. A great collective effort is necessary, which itself requires a vast information campaign. We are seeing results of the work done by such organizations as The National Foundation, the National Association of Parents of Retarded Children, The Advertising Council, the President's Committee on Mental Retardation, etc., on the national level.

On the local and state level a great deal needs to be done to create sufficient awareness, not only for those who work with handicapped children or plan programs for them, but also for our state legislators who control the state funds for programs.

The planning which is necessary for attaining a goal must bridge the gap between the "status quo" and the "status go", defining the mechanism of proceeding from the here and now to the here and then.

In drawing a circle one can start anywhere and reach all parts by proceeding in either direction. Where one starts in planning, whether with goals or needs, can be a matter of personal preference or related to the nature of the goal or the need. The important thing is to complete the process wherever one starts, checking and rechecking all the way.

Planning in a vacuum is no planning at all. We have to work with what we have. Attempts to develop services for spina bifida children in a vacuum can only result in isolated programs without community understanding, backing or support. So perhaps planning should start in centers like this one where we have already a core medical program. It may begin with the question of how many children there are in the area. What are their diagnostic classifications, their level of functioning? What is their age group? What needs are met? What needs are unmet? Which of the unmet needs have the greatest priority?

There are important fringe benefits or spin-offs from planning, and these are public education - a more informed citizenry, community organizations and some attempts on the part of agencies to communicate with each other and perhaps even to coordinate their activities.

Since the mentally and physically handicapped child needs the active aid of his parents, their attitude is crucial. No secondary emotional defect should be added to the original handicap; therefore, the earlier a frank but humane and understanding explanation is given to the parents, the better for all concerned. Well intentioned evasions of the truth or brutally candid pessimism can be disastrous. It is here that the organized services can help tremendously. Both emotional and material needs must be met through many varieties of direct services to make easier the management of the child as well as the rest of the family.

Regular cash allowances would enable any family with a severely handicapped child to purchase some necessary services such as drugs, household help, etc., which would be of assistance to the family. This takes place in many counties at this time; during the last legislative session such a bill was introduced in the Colorado State Legislature, but unfortunately it was not passed.

Adequate housing with ground floor accommodations is a necessity in the case where a mother has to carry a child up and down several flights of stairs each day in addition to her normal housework. The child in this situation is often made a virtual prisoner in the house and thus deprived of activity, experience and sensory stimulation which further retard his development.

When a mother comes home with a new baby, she is required to carry responsibilities for which she is totally unprepared; then it is of utmost importance to have a knowledgeable and perhaps specialized public health nurse or visiting nurse call upon her at frequent intervals. Considering the wide range of services needed by such families, a worker with special experience in the field should be employed as a coordinator for the provision of social, economic and health services to each such family in a given geographic area. The same coordinator would be knowledgeable regarding the gaps in community services for such families, and should act as a community organizer to work toward meeting unmet needs.

Studies of family problems in relation to congenital disorders have stressed the need for adequate genetic counseling. When a mother has had one child with spina bifida, the risk of recurrence is undoubtedly a serious one. Therefore, genetic counseling must be available to allow the parents to decide whether they want to avoid further pregnancies.

Reducing the mother's responsibilities by allowing the child to attend a pre-school program not only widens the child's experience but lessens the mother's fatigue. A tired mother is never a better mother. Here in Colorado the state legislature made funds available to communities to fund day programs from pre-school to adult sheltered workshops for mentally retarded and physically handicapped persons who are not accepted by the public schools. There are four such programs in the greater Denver area in Denver, Adams, Arapahoe and Jefferson Counties, administered through the Department of Institutions. These programs suffer from lack of funds and more work needs to be done to alleviate this situation and to allow these grass-root programs to grow.

Transportation must be provided, especially for those whose financial resources are very limited, so that they can secure the necessary treatment and other services. This is a major problem of expense to the community mental retardation programs and is a budget item in each program.

The services of a coordinator who would also provide information and referral services might also include an additional lifetime counseling program. Certainly the whole gamut of educational services is an important one. Whether these services are tendered under public school auspices or private ones isn't an issue now -- as long as these children receive the quality of services that they need.

Mental Health Services may be necessary for the child, the parents and/or family. Certainly we would hope that the generic mental health services would make their resources available.

An exposure to vocational activities should start early on the secondary school level and tie in with work-study experiences. These programs are financed through the Division of Vocational Rehabilitation which also provides the necessary counseling staff. For those who are unable to enter the competetive labor market or require extended vocational training, the services of sheltered workshops and activity centers are needed. At the present time, there are fourteen sheltered workshops in the greater Denver area. They are subsidized with funds from the Department of Institutions, the Division of Mental Retardation and formerly from the Division of Vocational Rehabilitation. Some receive direct Federal funds from the Division of Mental Retardation, Health, Education and Welfare.

For those who are not able to support themselves as adults there is Aid to the Disabled or if the patient is able to partially support himself, supplemental aid to the disabled is available. Social Security for the parents is another financial resource. If the child's father is on active duty in the military, there are commendable benefits for a child who is handicapped.

For those patients, who can live by themselves and to those who need the services of a halfway house here we need to be concerned about house parents, recreation and adult educational service. We must not lost sight that educating parents for their difficult work is not easy. The basis of their education must be individual and centered on their own child but as soon as they are ready they should be introduced to other parents of similarly handicapped children, preferably older than they are. This removes the feeling of isolation and allows a sharing of first hand experience. Parent groups often become the hub of a community activities dealing with handicapped children.

Important as the family is, it cannot replace society itself and today in a time of far-reaching changes, the public must be called upon to accept the handicapped. People who might wish to make a career or give voluntary services to the handicapped should be stimulated to do so. Industry should be urged to give them possibilities of doing useful work whether it is in their factories or by giving contracts to sheltered workshops. The Division of Vocational Rehabilitation which in this state is one of the main fiscal supporters of sheltered workshops must agree to provide funds which would benefit those handicapped who may not become productive vocationally.

As mentioned previously a coordinating mechanism, based upon an evaluation and treatment program should relate to the services existing in the community. Such a program would have the added advantage of offering a consultative service to the general or specialized agency; thus it would be more able to enlist the cooperation and use of its coordinative role by existing agencies. This would result in such needed side effects as an agreement on terminology, bringing improved communications, better data collection, and more unemotional and rational planning. The coordinating mechanism should be concerned with funding and eliminating those barriers which prevent our patients from access to community services.

More than that, this coordinating agency should serve in the coordination of in-service education and training and as a meeting ground for planning across agency lines. It might also be part of a community coordinating mechanism with a far wider role than merely the disability we are discussing here. If this is to be the case, however, it should have an identifiable subsection responsible for spina bifida. This approach promoted by the Council of Community Services was tried in Ogden, Utah, with commendable success.

The question of whether spina bifida patients should receive services in specialized or generic programs is an important one. By generic agency is meant any health, welfare, educational, rehabilitative, or employment agency in the community whose purposes are not for the specific care of any one disease entity. An example might be an orthopedic clinic not specifically for birth defect patients which would be considered a specialized service in other circumstances but would be considered generic in this definition.

Financially no community is able to have a wide range of specialized services. Available manpower also limits this activity. I am not proposing an either/or situation in regard to generic versus specialized services. Both are needed, or in this case, whenever we can use services, we should do so. All these are needed for a balanced program but they must be properly fitted into the overall community services structure.

When we talk about planning and services we eventually must talk about money and manpower for programs, realizing that what is recommended today may not be in force during the coming fiscal year, or even next month as a new program; nevertheless a review of those Federal grants which may be pertinent in the area of Service is worthwhile.

1. Project Grants for Health Services for Children and Youth via the Children's Bureau (Schools of Medicine and Teaching Hospitals are eligible)
2. Crippled Children's Demonstration Projects via the Children's Bureau.
3. Research and Demonstration in the Education of the Handicapped, Office of Education

4. Project Development Grants -- Rehabilitative Services Administration - basically a small planning grant
5. Mental Retardation Staffing Grants -- Divison of Mental Retardation and Rehabilitative Services Administration
6. Project Grants for the Rehabilitation of the Mentally Retarded - Division of Mental Retardation and Rehabilitation Services Administration
7. Mental Retardation Construction Grants
8. Research and Demonstration Grants - Rehabilitation Services Administration

There are also State Grants which go to the community programs for the retarded and physically handicapped to assist them in the operation of Pre-School, School and post-School Programs. Such funds are administered by the Department of Institutions and most important there is the state crippled children's programs which provide a variety of important services.

The problem of childhood crippling is a complicated one, requiring many approaches for its solution. There are at present a number of public and private agencies which are prepared to assist in both defining and solving problems, but the greatest deficiency lies in the area of manpower. Continued effort must be exerted to obtain voluntary and public funds to generate research and training of professional people.

MATERNAL ATTITUDES AND BEHAVIOR AS RELATED TO THE SELF-CONCEPT AND BEHAVIOR ADJUSTMENT OF CHILDREN WITH BIRTH DEFECTS

Claire Fishman, Ph.D.

I'd like to talk with you today about the research project we have been conducting here at the Birth Defect Center, about some of the ideas behind the project, about the procedures used and about some of the hypotheses we are investigating. The overall purpose of the project is to systematically explore some of the maternal attitudes and behavior which are associated with the self-concept and behavioral adjustment of children with birth defects.

Over the past few years there has been an increase in quantitative and systematic studies into the emotional adjustment of children with birth defects. Most of these studies have focused on the comparison of various groups of handicapped children with normal children on different aspects of adjustment. Very few studies have investigated differences among handicapped children. We know from clinical experience that some of these children are remarkably well adjusted to their handicap and to their daily experiences. They seem to have arrived at an effective and and well-balanced self-picture which includes their handicap as well as their other personal characteristics. Others seem to be completely overwhelmed by their handicap; they have great difficulty in coping with their day-to-day experiences and are unable to assimilate their defect into an integrated and positive self-picture.

How do these differences arise? And what can we in the helping professions do to promote and facilitate the healthiest adjustment possible under the very stressful conditions created by the defect; what can we do to promote and facilitate a more healthy environment for the growing child with a birth defect? At this point, we don't really know the answers to these questions. There are lots of ideas and lots of folklore based on many years of clinical experience, but few well-designed systematic studies to scientifically evaluate these ideas and folklore. We are making a small but hopefully meaningful start in this direction by systematically and quantitatively investigating some of the maternal attitudes and behaviors which seem to beat least partial determinants of the disabled child's self-concept, specifically, and behavioral adjustment generally.

Our emphasis on the child's self-concept is indicated by the critical and central role that self-concept plays in an individual's total personality and overall adjustment. And it would seem crucial, in the interests of primary prevention, to uncover the specific parental attitudes and behaviors which are associated with the self-concept of these handicapped children.

Subjects

The overall design of the research called for the individual assessment of 40 children, 20 boys and 20 girls, and their mothers. Ideally, our subjects would have been limited to a narrow, perhaps 2-year age range, to certain specific disabilities, and to a homogeneous family background. Great variations in type and severity of defect, as well as in age and social background, could hide some real and important patterns of relationships between maternal and child variables. However, in order to obtain enough subjects, we had to accede to the realities of the situation and take almost any type and severity of defect and accept a wider age range. Specifically, the following selection criteria were established: the children had to be Caucasian, between the ages of 8 and 14, have an IQ of at least 80, living with their natural mothers, and sufficiently able to hear, see,and speak clearly so as to permit adequate response to the various measurement procedures and adequate communication with the examiner.

Previous research in which mothers were asked to voluntarily participate have frequently yielded rather poor rates of participation, so we put a great deal of thought and energy into our procedures for our initial contacts with mothers. We were most gratified by the positive payoff of our efforts: of a total of 44 mothers contacted after a preliminary screening on our criteria, all but four agreed to participate. This is very important, since a substantial number of refusals to participate is a headache for any research: it obviously means a less than random sampling of subjects; and in our study, perhaps a restriction to only the best and healthiest of family situations. Of the 40 subjects seen in the project, 5 had to be excluded for not fitting in with the selection criteria. Of the 20 remaining boys and 15 remaining girls, 26 have had at least some contact with the Birth Defects Center; the others were obtained through individual contacts with non-clinic physicians in the Denver area.

Procedures

To avoid any possibility of bias, two different people were involved in the assessment procedures. A research assistant was trained to interview and administer all of the procedures to the mothers; she had no prior contact with the child nor any prior access to information about the child's personality and adjustment. I interviewed and administered all the procedures to the children and had no prior contact with or access to any information about the mother.

Let me briefly outline the procedures for both mother and child. I should mention first that all the sessions were tape recorded and transcribed, following the suggestions of several authors who bemoaned the loss of valuable data as a result of not recording.

First, the procedures for the child, in order they were administered. We borrowed a technique developed by Richardson and used by him in a study comparing handicapped with normal children. The technique is, called, somewhat inelegantly, the "Tell me about yourself" technique.

After a short warming-up period, during which the examiner (myself) did most of the talking to help the child feel comfortable, the child was asked simply to "Tell me a little about yourself, whatever you'd like to tell me, so that I can better get to know you". A majority of the children, regardless of age and IQ, responded readily. Following this, the child was interviewed for about an hour on a variety of areas. He was first asked about aspects of his life not directly involving his handicap: what he liked best and least about himself; what his day-to-day activities were like, specifically, the activities of the last typical school day prior to his visit with me; had he ever felt sad, angry, afraid, and proud and, if yes, to elaborate on that experience; what are his future plans and expectations, covering such areas as education, occupation, marriage and family, and hobbies. Next the child was asked why he had to come to the clinic, or to the particular physician who followed him. The question was completely open-ended, with no mention by me of his birth defect. Some of the children immediately explained, in varying degrees of accuracy and detail, that they had a particular type of defect. There were others, however, who either did not know why, or did not wish to tell me anything more than that they come for "check-ups and things like that." For those who did mention the defect, additional questions followed, essentially allowing them to elaborate on what they knew about the condition. Following the interview, the child was told he could take a break, but first I wondered how he felt about all those many questions I had asked. This was done primarily to ease up any possible build up of tension and to allow the child to ask any questions he might have had. In fact, their comments provide very interesting data in themselves. At least half of the children could be described as enthusiastic, another quarter as acquiescing and agreeable, and the remaining as cooly non-committal.

After a soda-and-cookies break, a Draw-A-Person task was administered. This was followed by two previously well-validated paper-and-pencil questionaires assessing self-reported self-esteem and the extent to which this report was defensively distorted. The child then completed a procedure I developed in hopes of tapping his underlying sexual identification. A storytelling task was next used to elicit fantasy material about the specific issue of being handicapped; the specific stimuli included pictures of handicapped children in a variety of activities with non-handicapped persons and the child was asked to tell a story about the picture. Finally, as a short screening device, the WISC Vocabulary subtest was administered to obtain a gross estimate of Verbal IQ. The whole session lasted anywhere from 2 to 3½ hours: a long period, but the children for the most part did not seen to mind it.

From the specific procedures in the child's session we are able to develop variables which tap the child's behavioral adjustment, his self-esteem, and his self-concept with respect to general issues and with respect to his defect specifically. In addition, the process of the child's interaction with the examiner--his ability to develop a friendly relationship, his ability to express a range of feelings, his ability to become involved in and enjoy the various research tasks, and so forth--all provide important evidence about his overall adjustment.

In order to augment this information obtained in the child research session, we obtained physician's ratings as to the nature and severity of the child's physical problems, and school reports including IQ and achievement test scores, grades and quantitative teacher ratings of the child's adjustment in school.

The session with the mother also lasted anywhere from 2 to 3½ hours. It consisted of a detailed, semi-structured interview, which I will elaborate on shortly; a story-telling task, using specially designed pictures and a specially designed coding scheme to get at underlying maternal attitudes and a short form of the MMPI, called by its author the Mini-Mult. It is 78 items and far more palateable to subjects than the 576 item questionaire. Like its longer equivalent, it was developed as a gross empirical measure of psychopathology.

It is of interest at this point to mention some findings that emerged from a preliminary analysis of our data. We were concerned with the extent to which the child adjustment and self-esteem variables would be related to certain gross indicators of external stress, including the social class of the child's parents, the extent of marital stability in the child's family, the extent of the mother's general psychopathology, the severity of the child's physical problems and handicap and the child's IQ. Statistical analysis of our data revealed very little if any relationship between these gross external stress variables and the child adjustment variables. This would seem to suggest that there are more subtle determinants of a child's ability to adapt to his physical problems than the gross degree of external stress he experiences. From a prevention point of view, this finding appears quite hopeful, for these gross stress variables that I've mentioned such as severity of defect, are generally very difficult, if not impossible to alter; the maternal variables that are functional in this situation might well be quite amenable to intervention.

Hypotheses and Preliminary Results

The relevant maternal variables in the study were developed after an extensive review of the literature and are mainly assessed by a detailed coding and rating scheme of the semi-structured interview with the mother. To give an idea of these variables and their associated rationale, let me review with you in some detail a few of the specific questions in the interview. We begin by asking about the history of the situation: when and how the mother first learned of the child's defect, her interaction with her husband and other family members concerning this information and her specific reactions to the information, including her feelings and actions taken, her understanding of the nature and cause of defect, and her expectations about the future. Obviously, the answers to these questions are retrospective and are subject to distortion. However, in the absence of extensive and expensive longitudinal procedures, a retrospective measure is frequently employed as a way of estimating the past events.

Basically, these first questions are aimed at finding out about the way the mother coped both affectively and cognitively with the crisis of giving birth to a child with a defect. In the folklore, there are two opposing viewpoints regarding the healthiest way to handle a crisis such as the birth of a defective child. One view holds that it is best for the mother to keep a stiff upper lip, that it is best for her to keep busy so as to keep the very upsetting feelings out of her mind, and that she should "stop worrying since there is nothing you can do about it anyway". Someone who behaves this way, or who, in a fatalistic manner, seems to accept such a tragedy as God's will , is often upheld as a model of strength, psychological health, and even moral virtue. The opposing position, propounded energetically by such psychiatrists as Gerald Kaplan and Eric Lindemann, essentially claims that an individual's active emotional and intellectual confrontation of a tragic event is essential to a successful resolution of the inevitable grief reaction that will accompany the event. That is, in response to tragedy, the individual must fully experience the reactive negative emotions, he must seek out information regarding the event, and he must seek out and receive emotional support from significant others in his environment. In terms of the tragedy of having a defective child, the "stiff upper lip" view is often associated with the wish to protect the new mother from as many of the frightful truths of the situation as possible to minimize the difficulties and danger, allowing her the opportunity to experience the joys of childbirth free of fears and despair. This facilitates a healthier climate for the mother and, perhaps more important, permits a more normal realtionship to develop between the mother and child.

The "crisis confrontation" view is often associated with the belief that early confrontation is essential, that frankness with the mother is critical since the mother's inevitable and unverbalized suspicions can lead to very distorted, non-genuine interactions with her child. Another danger according to this view, is that the mother is in for a terrible disappointment at some point, and it is harder for her to constructively cope with this disappointment at a later point once a different kind of relationship with the child has already been established. This is really what we are interested in in the final analysis: the quality of the relationship between the mother and child. And we hope to explore this whole question of initial confrontation by the mother as it appears to be associated with the later adjustment and self-concept of the child.

Turning to the next area dealt with in the mother her present beliefs and feelings about the child's disability. Having a child with a birth defect is often an ongoing crisis; the crisis doesn't end with the weeks or even months after the newborn child goes home from the hospital. The child going to school for the first time, the child coming home crying because of cruel teasing, the child requiring surgery which will transform his body to a significant degree, the child going to the mother and crying as did one of the spina bifida children in the project: "Mommy, I'm not a human because I can't go the bathroom like a human. Why did God pick on me?" All of these events daily beset the parents of these children, and along with the continuing threat of illness and death, constitute a state of perpeptual crisis. And so, the material I've just discussed on the mother's early reactions are all relevant here. How is the mother coping with the present situation in terms of emotional and intellectual confrontation. Again, we have two opposing possibilities with, of course, all gradations in between: on the one hand, the mother who manifestly shows calm and minimal concern about acquiring additional information. And on the other hand, the mother who is openly emotionally upset by particular events and who continually demands information and clarity, even perhaps where it is not possible. This is the mother who frequently irritates staff members. Aside from what is best for the staff, what would seem best for the child? An interesting finding that seems to be emerging from the data is that the mothers who are both frequently emotionally upset and who are most aggressively demanding of information and action concerning their child have children who are among the best adjusted in our sample, on the basis of school reports and my own observations. However, this finding is still preliminary and impressionistic and must be further documented through a more rigorous analysis of the data.

Another series of questions was designed to assess the mother's image of the child, paralleling our assessment of the child's image of himself. We simply start out by asking the mother, "How would you describe your child so that a person like myself who doesn't know him would get an idea of what kind of child he is?" We also ask the mother to describe the characteristics she likes best and least about her child. There are several specific variables derived from this area: how differentiated is the mother's picture of her child, that is, to what degree does the mother have a complex, variegated picture of the child, with separate physical and psychological components, with both positive and negative features, and so forth? How realistic are her expectations for her child? How much understanding does she have of the child's disability? How much has the mother worked through her negative feelings over her loss, that is, how available are her negative feelings regarding the loss and how appropriate are they? Similar variables can be derived in terms of the child's self-image: how differentiated are his self and body images? How realistic are his expectations for the future? How much understanding does he have about his condition? And how available and appropriate are his negative feelings about his defect? We are hypothesizing that there is a relationship between these various aspects of the mother's concept of the child and the child's self-concept, and that these aspects of the child's self-concept are in turn related to the child's self-esteem and adjustment. An alternative hypothesis, one frequently suggested or implied in writings on the matter, is that the basic emotion of love and undifferentiated acceptance by the mother of the child is the critical variable associated with the child's adjustment. Moreover, in this view, the idea is frequently expressed that self-acceptance is no great problem among birth defective children. For example, in a Newsweek article on thalidomide children, March 4, 1968, a physician specializing in treating limbless children, was quoted as saying: "They accept themselves easily. Unlike a child who loses an arm after nine or ten years of life, they have never known normality, so they are not conscious of the defect."

In contrast, our hypothesis is that it takes a great deal of psychological work on both the mother's part and the child's to achieve a well-balanced, well-integrated, well-esteemed picture of the child; and that the success of the mother in performing this work is critical to the child's ability to perform it. Thus, not before both the mother and the child perform this work can success be achieved.

The last area I'd like to describe to you involves communication between mother and child, specifically with respect to the defect. We begin this section by asking the mother: "I wonder how your child feels about the fact that he has this condition?" And we follow this up with specific questions about the extent to which the two have talked about the condition and associated issues. We also ask the mother about her general attitude regarding communication. Again we might refer to two opposing views on this issue: some people feel that there's enough discussion and enough emphasis placed on the defect from all the medical work-ups and from the sheer time and effort involved in the physical care of the child; and that it would be best to eliminate as much as possible any discussion of the defect so as to deemphasize it as much as possible. On the other hand is the view that there's no way of reducing the impact and stress of the defect by a "web of silence", and that keeping down the discussion only serves to emphasize the degree of concern and distress with the message: it's too awful to talk about. Somewhere in the middle is the view that if the child brings up the issue, then it's appropriate to engage in a discussion about the condition.

It is our hypothesis that open discussion of the defect and associated issues is essential to the child's self-acceptance and overall adjustment. Open discussion first communicates to the child that his defect is after all a permissible subject of conversation and neither too shameful, nor too frightening, nor too ugly to talk about. Second, it provides the child with an opportunity to air out his otherwise unexpressed fears and frustrations. Unexpressed, these fears and frustrations can assume overwhelming proportions, way beyond the reality situation. And even where the fears and frustrations are reality-oriented, talking about them can lead to better understanding and more adaptive coping behavior. The frequently suggested alternative of allowing the child to take the lead and determine what and how much ought to be discussed implies that the child automatically feels comfortable, or at least can overcome his feelings of discomfort about the condition, enough to initiate discussion of underlying thoughts and feelings. It is my strong impression from my experience with these children that this cannot be assumed. Not all parents feel comfortable about openly discussing these issues with their child, and obviously discussion under such conditions would be uncomfortable for the child too and defeats its purpose. If our findings suggest that this variable is indeed an important one, work with the mother to promote open and frank attitudes and behavior would seem not only appropriate but critical from the point of view of primary prevention.

In conclusion, I'd like to say a few words about early intervention into and primary prevention of adjustment problems of birth defective children, which after all is one of the main purposes of this research project and other projects like this. A very important focus of concern in the helping professions should be early intervention and primary prevention: attempting to prevent potentially hazardous conditions from arising and creating more pathological conditions by modifying and encouraging more healthy environments in the first place. All too often, in the squeeze of economy and manpower, this primary prevention and early intervention orientation is sacrificed to the most immediate crisis demands, and the potentially hazardous conditions have a chance to develop and set the stage for later, more intractible psychopathology. And then, of course, more complex, more costly, techniques of intervention--tertiary prevention Gerald Kaplan calls it--must be set in motion, techniques placing far greater demands on manpower.

It seems to me that research of the kind I've just described has not only theoretical implications, but also very practical implications in this regard. Once we uncover some of the variables which are critical for the well-being of the birth defective child, it will be not only possible but imperative that programs be instituted which are designed around specific issues and with very specific goals. These programs can be designed for each level of prevention. At the level of primary prevention, specific ideas based on well-replicated findings could be publicly disseminated. For example, we have found that all the children in our research suffer from cruel teasing by school mates, and this would definitely seem to have very practical implications for educational programs in the schools attended by these children. To date there has been no systematic attempt to encourage such programs in the schools, but from my contacts with many very interested school principals, it seems clear that many would welcome specific recommendations from us.

At the level of secondary prevention, procedures such as the ones used in our project, could be used to screen out high psychological risk groups for specific intervention programs. Thus, if poor communication between mother and child proves to be a key determinant of the child's poor adjustment and self-concept, mothers who manifest difficulties in these areas could be enlisted into programs specifically designed around this issue. Behavior therapy, for example, has been found to be very successful in the rapid modification of attitudes and feelings preventing open communication. Finally, at the level of tertiary prevention, where full-blown problems are found to exist, and there was no shortage of full-blown problems among the children in our sample, research of this type can help direct our attention to the more likely factors underlying the psychopathology.

MANAGMENT

COMPREHENSIVE MANAGEMENT OF THE NEWBORN WITH SPINA BIFIDA

THE ROLE OF THE PEDIATRICIAN

Janet M. Stewart, M.D.

The complexity and multiplicity of problems associated with meningomyelocele are well known and the importance of a multidisciplinary team approach to these problems has been well accepted. Most medical centers offering care for these children have established a meningomyelocele clinic or "team" consisting of pediatricians, social workers, nurses, neurosurgeons, orthopedists, physiatrists, urologists, physical therapists and psychologists. Eventually most children are referred to such centers for their long term follow-up care.

It is important that this same approach be utilized in the care of the newborn with a meningomyelocele. The birth of a child with such a serious and life-threatening defect is frightening and bewildering to the parents. From the onset, they need the best in coordinated medical care with careful explanations of the significance of the defect, the kinds of problems that can be anticipated and the type of therapy that their child will receive. The medical approach to the newborn may vary from center to center, but there are certain basic principles which are essential for the achievement of optimal medical care.

The infant with a meningomyelocele should be transferred immediately after birth regardless of the severity of the lesion to the center selected for definitive care. Too often the baby is "left to die" but a week later with the infant alive and infected, the parents are confused and bewildered. Although the treatment for each child may vary, the decision should be in the hands of those who will be caring for the child as soon as possible after birth.

The child with meningomyelocele should be admitted to a joint pediatric/neurosurgical service or to the pediatric service with close neurosurgical association. It is apparent that the immediate problems and decisions are neurosurgical, but the pediatrician has much to offer in the areas of fluid and electrolyte balance and antibiotic therapy. Then too, a significant number of these children may have associated anomalies which need to be recognized and treated.

The problems of the child with spina bifida, even in the newborn period, are not exclusively neurosurgical and each member of the team should be involved in early evaluation and treatment and baseline urologic studies are necessary. Urinary tract infections which may occur shortly after birth must be appropriately treated. A careful appraisal of muscle function should be done for several reasons: to assess and compare early with ultimate function, thus getting a more objective appraisal of the effectiveness of therapy; to start the family on a home program in order to prevent contractures and to stimulate normal development. This involves the orthopedist, the physiatrist and the physical therapist. The social and financial implications of this malformation are tremendous and the social worker very early becomes a major source of support for these families.

The very nature of a meningomyelocele makes contact with various medical and paramedical subspecialists necessary, but with this contact may come conflicting opinions and confusing advice. It is therefore essential that there be one primary physician who is the principal source of information and support for the family. He should begin this association as soon as possible after birth. This physician is most often the pediatrician who is perhaps best trained to answer the many broad-based questions of the parents. It is not his position to make neurosurgical or urologic decisions but rather to interpret them to the parents and to fit them into the broader context of the child's total therapy program. There may well be differences of opinion as to the particular aspects of care. This disagreement is healthy for the medical staff but devastating for the parents. They should not bear the brunt of medical ignorance and confusion. They need to be told the truth and the limitations of what can be done, but conflicting stories from each subspecialist will not help the parents or the child. The primary physician should be the major source of information. He may need to arrange conferences between the parents and the surgeons, but he should be present to anticipate confusion and to help answer questions for the parents.

The primary physician may encounter problems in several areas: the house staff may not be familiar with this concept of care or may not recognize its value, and since most centers are in teaching hospitals, house staff will be involved in the care of these children. The problem of spina bifida is long term, and the family needs prolonged medical and social service contact. They do not benefit from seeing a different physician each time they visit the hospital. The importance of one physician who can constantly be a source of support and information for the parents is most important.

There will also be problems in answering the questions which the parents bring. They have often been told many things in the brief time between the birth of the infant and the time of transfer. They have been told that there is "no hope" and that they had best institutionalize the child. The facilities for this type of care are extremely limited in most states. Even if this were the best solution, it would be impossible in most cases.

The parents quite naturally want to know what the future holds for their child and their questions may be difficult or even impossible to answer. It may be possible to anticipate potential motor function and bowel and bladder problems, but the prediction of intelligence is much more difficult. The parents fear mental retardation and indeed there is an increased incidence of retardation in children with spina bifida. They can only be given the statistics, reasured that good medical care will increase the chances for good intellectual growth and promised help with their child as the years reveal his ultimate potential.

The primary physician very early becomes one of the main sources of support for the family . He attempts to answer their questions and eliminate any confusion in their minds. He coordinates the medical care for the child so that optimum care can be given with a minimum of time, discomfort and expense. He discusses fears for future pregnancies and gives or arranges genetic counseling. Most important of all, he works with the family through the years as they face each problem and helps them to accept their child for what he is and to make realistic plans for the future.

It is evident from a symposium such as this that comprehensive care is essential to the well being of a child with as complex a defect as a spina bifida. No one physician can do it alone. This comprehensive care should start in the newborn period and should be coordinated by one physician who can work with the child and the family throughout the long years of medical problems.

There is still controversy concerning the best way to medically manage a child with spina bifida. There is little argument, however, that guidance, education and support of the family play a major role in the achievement of optimum potential.

COMPREHENSIVE MANAGEMENT OF THE AFFECTED NEWBORN

The Role of the Social Worker

Margaret Mc Call, M.S.W.

Arranging for the comprehensive care of a newborn child with spina bifida is a most complex affair. Dr. Stewart has discussed problems in arranging medical care with the hospital. I would like to touch on some of the problems of working with parents on plans for post-hospital care and on the involvement of paramedical personnel in making these plans.

First and foremost any plans for a newborn child must be based on the physical and emotional resources of the family into which the child is born; these are usually fairly easy to evaluate. There is the need for supportive help of many kinds -- financial assistance in meeting the costs of medical care, nursing assistance in planning for other members of the family -- however, this discussion will attend chiefly the assistance needed in handling the emotional impact on a family when a child with spina bifida is born. These factors are not easily recognized, and they have a significant effect on the success of post discharge plans.

The birth of a defective child creates a severe emotional crisis for any family. The ways in which this is handled vary greatly, but for all families the problem is essentially the same. Parents must handle concurrently the tremendous disappointment of losing the normal healthy child that they had hoped for while adjusting to the birth of a threatening, anger-provoking child who represents their worst fears, rather than the hopes that have been developing during the preceding months.

Solnit and Stark made an excellent study of this adjustment process in their monograph entitled "Mourning and Birth of a Defective Child." David Kaplan develops a similar thesis in his article entitled "Maternal Reactions to Premature Birth Viewed as an Acute Emotional Disorder." Both of these studies are applicable to working with the families of children with spina bifida. These authors feel that it is necessary for the mother to recall and grieve over her longings for a normal child before she can liberate her feelings to develop a warm relationship with her defective child. They point out that the process of interpretation must be aimed at balancing the reality of the handicapped child's condition against the parent's pre-existing hopes and the fearful fantasies that most mothers have experienced during their pregnancies.

Ideally this kind of interpretation extends over several interviews to allow the initial disbelief, denial and distortion to work into awareness of disappointment. At times both parents and medical personnel will be tempted to avoid this confrontation by seeking immediate placement of the defective child away from home. In some cases placement is an appropriate treatment plan, but unless it is approached with a clear understanding of the reasons for it, the parents may never have the opportunity to recover from the blow of having produced a defective child.

In most cases the newborn baby with spina bifida remains in the hospital long enough to allow for several interviews; but too often these interviews are focused entirely on plans for medical treatment. Parent's initial reaction to the crisis may obscure the underlying emotions; those who visit regularly and show concern about their child but not excessive depression may not be handling their feelings well. Visiting is a poor indicator of a mother's feelings for her child. Allowance for the mourning process may cause some parents to stay away to work through their disappointments; they are then more ready to participate in realistic planning than the parents who present a facade of acceptance. With staff limitations we seldom have time to contact parents outside the hospital, and we must be aware that relationships between families and medical personnel within the hospital are affected by the treatment atmosphere.

The baby with spina bifida is usually placed in a special treatment ward such as a premature center, which has an atmosphere of serious concern about serious medical problems. This environment contrasts markedly with that of success and pleasure which often pervades a regular maternity ward. This means that although parents are encouraged to visit as much as possible, they are still outsiders and have little opportunity to develop their role as parents. Following a normal delivery,a mother usually begins to develop feelings of competence very quickly. If she is able to respond to her child's needs, this provides a basis for the mother-child relationship. Pride and warmth of feeling go hand-in-hand with success. When a handicapped child is born, everyone becomes apprehensive in their contacts with the mother. In the case of the child with spina bifida even the baby's survival is in question and the mother seldom receives congratulations for a job well done. With continued hospitalizations, fears of inadequacy as a mother are intensified and unwanted dependence on medical personnel may be expressed in avoidance of hostility.

We can hardly expect parents to be fully appreciative of our efforts to help their child when the whole situation of needing this help was unwanted and unexpected. The feeling, sometimes openly expressed, that things would have been much better if the child had never been born really should be expected and handled as openly as possible. Fathers manifesting hostile concern about the costs of medical care should also be expected. The handicapped child is quite a threat to a man's image of himself as a provider for his family as well as to his self-image as a reproductive man.

To demonstrate the intricacies of medical treatment planning, two families have been selected who have been known to our Spina Bifida Clinic staff during the past year. As you will see, the treatment planning process differed markedly according to the emotional resources of each family.

The first case is that of baby girl Patricia born August 23, 1969, the first of twin girls, the result of Mrs. N's first pregnancy. The other twin is a normal healthy child who was discharged from the hospital with her mother while Patricia was transferred to Colorado General Hospital immediately for surgical treatment of a meningomyelocele. She also required casting for equinovarus and insertion of a shunt for hydrocephalus. She did well post-operatively and was ready for discharge at about 6 weeks of age.

Mr. N.Patricia's father, contacted the Birth Defects Center shortly after her admission requesting an interview to learn more about spina bifida. At the time of his initial interview he said that he had been told that some children with spina bifida were placed in foster homes if their parents were not in a position to care for them at home, and he asked for specific information on arranging a foster home placement. The private pediatrician called me shortly after this initial interview to say that she did not think that these parents should be expected to take Patricia home.

From the very beginning there was pressure to make a discharge plan as soon as possible although it was still impossible to predict what Patricia's medical needs would be. Mr. N.was completing his doctoral dissertation and originally planned to leave Colorado in early October to take a post-doctoral position outside the United States. He stressed that this post-doctoral position was vital to his professional future. During our interview, it was learned that the N's had known each other less than two years. Mr. N. mentioned that both of them had had periods of depression in the past and that meeting and marrying had been vital in alleviating their depression. The N.'s seemed to have wanted a child at this time but they were very concerned about their financial stability and the appearance of twins was in itself threatening to these concerns. Their plans for leaving the country made it very difficult to offer them concrete assistance in meeting the ongoing costs of Patricia's care.

Because the N.'s were on the verge of departure -- they actually postponed leaving for a month after Patricia's discharge -- the discharge plans were made in a series of four interviews -- two with Mr. N, alternated with two joint interviews. I saw them alone for the first two sessions. In his individual sessions, Mr. N. appeared to be trying to spare his wife the pain of discussions of plans for Patricia or to assume a leadership role, but as our discussions moved into more depth of feeling he became aware that he could not be solely responsible for such major decisions.

Both Mr. and Mrs. N. came to the initial interviews with the idea that foster home placement might be the best arrangement -- admitting that they really didn't know anything about foster homes. We reviewed Patricia's condition and their own situation, emphasizing that Patricia's prognosis was still uncertain. I tried to review the many possible variations in her future development. The conversations that developed during our interviews revealed two very different reactions to the birth of a handicapped child. Both Mr. and Mrs. N. based their plan of placing Patricia in a foster home on the idea that she would completely disrupt the picture they had developed of their future family life. Although they did not want an irrevocable placement, they were considering it in terms of at least two years which we thought would be disastrous to developing any feeling for Patricia as their child. They emphasized their interest in sports especially skiing, and pointed out it would be very difficult if one twin were able to joint with them and the other were not. They both pointed out that the financial drain of Patricia's care would interferewith the things they wanted to do and might lead to resentment of her. Philosophically they explained that they did not believe that a child had a personality until some time after birth. This idea seemed to relieve them of their commitment to this child.

Their ability to develop a relationship to the normal child also gave me the hope that once they saw Patricia as a real living being they would relate to her. This recognition of her as a person was of course hampered by Patricia's isolation in the premature nursery. Although there is hardly any restriction in visiting by the hospital, Mrs. N. was involved in caring for the other baby 30 miles from Denver and was not a frequent visitor. In spite of their sincere efforts to be unified in their own thinking -- they were both concerned about the threat to the stability of their marriage if they brought Patricia home and Mrs. N. became excessively involved in her care -- the N's reacted to two rather different trains of thought. For Mr. N. the breakthrough in his thinking seemed to come as we made reference to his picture of himself as a father, the fact that he would always be concerned about and be aware of this child. On a conscious level he maintained that placement might be for Patricia's best interests so that she would not be in competition with her sister, even if she were only moderately handicapped. When we focused on the normal twin, already very much a part of the family, he became aware that it would be difficult to justify to her the rejection of her sister. This appeared to be the point at which Mr. N. turned his attention to making plans to bring Patricia home.

Mrs. N.'s questions and conversation focused more on what Patricia might be like if she were significantly handicapped - her abilities, and her emotional reactions. It was a surprise to her that a defective baby could develop into a feeling person enjoying many normal activities. It was as if she were suddely able to mobolize some of her previous dreams and expectations of a girl child in place of what must have been her greatest fears. Both parents asked questions about every possible detail of Patricia's condition, which according to Dr. Fishman's study would indicate good adjustment in the future.

Following the fourth interview, Mr. and Mrs. N. decided to take Patricia with them, taking the chance that there would be financial assistance for Patricia's medical care, although she would not be eligible for governmental assistance. They stayed in Colorado about a month after Patricia's discharge. She did very well medically and her parents made frequent proud reports of her socialization with her twin sister. The question of foster placement was dropped without further discussion.

I should add that Mrs. N. welcomed the assistance of her public health nurse after Patricia's discharge. She kept in close contact with our staff by telephone and during weekly visits to the Orthopedic Clinic and the Department of Physical Medicine and Rehabilitation.

I am fully aware that this family may face many adjustment problems in the future since living with a handicapped child is a continuing problem of adjustment; however, I do feel that they have made significant progress in working through the mourning process, and in perceiving their physically imperfect child as a person. They also demonstrated their ability to request and make use of supportive help in a period of crisis. It is of utmost importance that medical treatment centers for handicapped children provide the personnel to meet these needs.

The second child I would like to discuss was born at a local hospital April, 1969, with a meningomyelocele, developing hydrocephalus and an enlarged heart. She was immediately transferred to Colorado General Hospital where she was treated surgically with spinal closure and the insertion of a shunt. Dispositional planning for this child then extended over a period of six months for reasons to be explained.

Angela was the second child of a 19-year-old Spanish-American mother and a 21-year-old Negro father. Her older sister now two years old had been born with a cleft palate. The palate had been successfully repaired prior to Angela's birth; however, the mother who was only 17 years old when this child was born, had reportedly been very upset by the child's handicap.

During Angela's initial hospitalization her parents separated as they had several times in the past and within the next few months Mrs. L. filed for legal separation and applied for ADC. Apparently Mr. L. made no contest for custody of the children and our contacts in making health care plans were entirely with the mother. During the six months of planning Mrs. L. moved back and forth between Denver and her parent's home, 127 miles from Denver.

Hospital discharge was first attempted in May when Angela seemed to be doing fairly well post-operatively. During the month of hospitalization Dr. Stewart and I had made several unsuccessful attempts to meet with Mrs.L. to talk about Angela. She visited seldom and always in the evening. Contact was finally made by one of our other social workers just before discharge.

The first discharge lasted three days until Mrs. L. brought Angela back to the hospital reporting excessive vomiting. She was readmitted for 15 days and again discharged to her mother. Two days later she was readmitted with the same complaint. Again she was carefully observed and no physiological cause for the vomiting was found. Problems in the mother-child relationship were thought to be a factor in the vomiting; however, on the ward she continued to vomit and was an irritable baby.

She was discharged to her mother for the third time on July 10th and remained home for about one month. During this time her mother brought her frequently to the Child Care Clinic because of excessive vomiting. The social worker saw her at most of her visits trying to let her express her feelings and concerns about this baby. A public health nurse made regular visits to the home. Mrs. L. did recognize that her own tensions might be affecting the baby but she was unwilling to take any medication or seek help for herself.

On July 11th Angela, now 3 months of age, was readmitted to the hospital weighing no more than her birth weight, and a referral was made to Denver Child Welfare requesting assistance in arranging for foster home placement. The next few months until her discharge to a foster home placement on October 27, 1969, were fraught with continuing medical problems including cardiac surgery.

For Mrs. L. it was a struggle to even communicate with professional personnel. She acted out this struggle by infrequent visits to the hospital and with passive responses when physicians and the social worker spoke with her directly. Verbally she expressed her ambivalent feelings for Angela saying "Of course I want to care for my child but I don't know how I'll ever be able to do it." This was a pretty clear statement of her predicament.

For the welfare department and for some of the physicians it was a struggle with their conviction that this mother should be given the opportunity to develop a satisfying mother-child relationship, which she would not have if separated from her child. Unfortunately some of the conflict and confusion among professional people about the plans for Angela carried over to conferences with Mrs. L.; however, it was ultimately decided that the emotional reserves of this young mother were only adequate to handle the problems of one handicapped child and Angela was placed in a foster home. I hope I am right in thinking that we made it possible for Mrs. L. to participate sufficiently in making these plans to feel some satisfaction in her performance as a mother. Angela is doing well in the foster home and it may be that when her medical problems are more stabilized, her mother will be able to care for her at home.

There is great need for material support for the parents of children with spina bifida during the newborn period, and relief for the financial burdens of major surgery and extended hospitalizations is essential. Families need assurance that help is available so that they can give attention to their emotional reactions.

Parents must have professional time beyond that needed to deal with medical and financial problems. Unfortunately we seldom have time to really understand the particular grief process a family is experiencing or to go out to meet them whenever they are comfortable to talk whether it be at home or in a professional office.

Convalescent homes for infants are needed to give families a chance to mobilize their resources. Our present system of trying to utilize welfare foster homes is very cumbersome. Because the welfare department believes that they should evaluate the situation and make their own decision on the use of foster homes, contacts with the family are duplicated and prolonged and the decision making process even with the best communication is transferred away from the medical center.

Finally, I would speak out against any consideration of early institutional placement for these children. No one should be placed in the position of trying to predict a spina bifida child's future during the newborn period.

The variety of medical specialities involved in a spina bifida treatment program demonstrates to some degree the complexity of treatment planning. In my remarks I have tried to show how comprehensive care is not complete without coordinated services to meet the social and psychological needs of families who are faced with the unexpected birth of a severely handicapped child.

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IMMEDIATE REPAIR OF SPINA BIFIDA

ETHICS AND POLICY

W.J.W. Sharrard, M.D., F.R.C.S.

Opinions concerning the timing of operations for meningomyelocele have varied over the years. Sometimes the spinal lesion has been operated upon in the neonatal period though not necessarily on the first day of life, simply to make the child more socially acceptable. In other centers, the operation was postponed until about 18 months because it was thought that the condition of the child would then have stabilized, any hydrocephalus would have developed and that there might be more skin available for closure. In other centers, operation was deferred in the belief and sometimes hope, that the child would die.

Such policies were acceptable in the past when, in fact, most children with meningomyelocele did die. Eighty per cent of children with meningomyelocele also had hydrocephalus and in 50 per cent, without treatment for the hydrocephalus, death was certain. Without antibiotics, the incidence of meningitis during the first week of life was extremely high. In time past, whatever was done, mortality was likely to be of the order of 94 per cent. It is since the advent of antibiotics and the discovery of adequate means of treating hydrocephalus that thought has had to be given afresh to a policy to be adopted with regard to the lesion of the spine. Before discussing details of this, it is necessary to consider what ethical policy one is going to take in the management of such children.

There are those who feel that such babies ought not to be treated at all because there are too many babies in the world already. Those who hold these views are really saying that they feel that the patient should die. Unfortunately, if these children are fed and nursed in the usual way and possibly given antibiotics, about one third of them will survive, and will present a problem for treatment and then for decision as to whether the hydrocephalus should be treated. To those who think that such children should die I believe that it is right that they should be the ones to ensure that the baby does do so rather than survive in a miserable state.

Our policy, that is the policy adopted by Mr. Zachery, Dr. Lorber and myself at the Children's Hospital, has been that, unless the child is obviously suffering from another lesion that is manifestly fatal such as a serious cardiac defect, it is right that he should be given every opportunity to live with the maximum of possible preservation of function.

TYPES OF LESIONS

In general, there are three main types of serious spina bifida. Meningomyelocele is a lesion, usually covered with skin and bulging at birth, which contains no nervous tissue. If it is a true meningocele and at operation no nervous tissue is found in the sac, excision of the sac alone should give a complete return of normal neurological function to the lower limbs. Meningocele occurs in about 7 per cent of this type of case. Meningomyelocele covered with skin with or without a lipomatous attachment to the cord or cauda equina is the second commonest lesion. It does not necessarily need operating on immediately after birth because the child is in no danger of immediate loss of nervous tissue function by infection or drying. We believe that it should be operated on at any rate within the first six months of life because of the strong liability to subsequent deterioration.

The commonest and most serious lesion is open meningomyelocele. This is most commonly found in the lumbar or lumbosacral region but it can extend for varying lengths of the spine. It is sometimes called rachischisis or myelocele but all lesions are really the same. A small number of meningomyelocele lesions have an open spinal cord but all the nerve cells in it are present and there is potential for normal function in the lower limbs and bladder. This probably applies to about 15 per cent. In the remainder, there are varying degrees of myelodysplasia in the open spinal cord and this may take various forms of upper or lower motor neurone loss or both.

IMMEDIATE CLOSURE VERSUS DELAYED CLOSURE

In 1963 a trial of operative closures of the spinal lesion within the first 48 hours of life was made. The numbers were small but the results showed clearly that mortality was not increased and that function in the lower limbs was very definitely better in the group that had been operated upon early. It is not so much that improvement in function may necessarily occur in the lower limbs compared with the state of the limbs two or three hours after birth but that, if the lesion is not operated on, there is liable to be a steady deterioration in the condition of the paralysis of the lower limbs. This is almost certainly caused by a combination of infection, drying of the plaque and swelling of the lesion causing a traction on the roots of the cauda equina. The results in the small series followed up to five years after the initiation of the trial showed that a few children who survived in the conservative group are all completely paralysed in the lower limbs whereas those who were in the operated group have a substantial amount of muscle presence and all of them are now able to walk independently.

In a larger retrospective series, the position as regards paralysis became even more clear. Since 1961 all children with open meningomyelocele have been operated upon within 24 hours of birth, however severe the defect. The results have shown that, at the third year of life, 50 per cent of children are surviving and, in recent years, I think the survival rate has become even higher, up to 75 per cent. Of those treated very early, 15 per cent had no significant paralysis either of bladder or the lower limbs. A further 19 per cent have slight paralysis of the intrinsic muscles of the feet and paralysis of the bladder. Forty-two per cent are partially paralysed but with sufficient muscle to be able to make use of orthopedic management and become easily independent in walking. Twenty-four per cent still have either moderately severe paralysis or paralysis with spasticity but even these can become independent in walking. This compares with those treated conservatively in which 12 per cent have nearly normal lower limbs, 28 per cent have partial paralysis, 60 per cent complete or severe paralysis.

It was also interesting to compare the results of immediate closure of large thoracolumbosacral lesions which have the worst prognosis when treated conservatively or operatively. Even in this group, the results of operation were better and there were even a few children who had completely normal lower limbs even though they had an open meningomyelocele from the mid-thoracic region to the sacrum. For this reason, there are no grounds for selection of patients with extensive lesions for conservative management, and everything points to the fact that all open meningomyeloceles, should be closed as soon as possible after birth.

Account has also been taken of the effect on hydrocephalus and we have found that the incidence of hydrocephalus in not altered by early closure. However, there are some patients in whom the hydrocephalus increases fairly rapidly after closure and there may be a case to be made out for treatment of the hydrocephalus within three or four days on closure of the spine. Normally, though, we would wait until three weeks before doing ventriculography to assess the degree of hydrocephalus and making a decision about the need for a Spitz-Holter valve, the reason for this being to try to avoid, as far as possible, any infection being drawn up into the central nervous system.

ASSESSMENT

When a child is admitted to the hospital, his general condition is assessed. It is essential that the baby should be warm. We have arranged that ambulances shall have special incubators available so that the baby's temperature will not become low. If a baby is operated on with a low temperature, it is very much more likely to die after operation. As soon as he is warm and the general condition is satisfactory, arrangements are made for operative treatment. Before the operation is begun, an assessment is made of the paralysis.

A muscle chart is made of the lower limb function. This is done, not so much in terms of the grading from 0-5, but simply of the presence of action or no action in the muscle groups in the lower limbs and the chart is made out in terms of level of root action. This very easily shows which patients have a root innervation down to a certain level with paralysis below this level. It is very difficult to be sure whether the movements that are present at birth are reflex or voluntary. In some cases there is definite spasticity present and there is no doubt about the upper motor neuron nature of the lesion; in others there is spontaneous activity which seems by every test that one can do to be a normal and voluntary use of the muscle. We also do a complete faradic stimulation of the muscles. We find that, if a muscle is working voluntarily and naturally, also responding to faradism, its chances of being present three months later is 90 per cent. If a muscle is not acting voluntarily but does respond to faradism, there is a 40 per cent chance that is will be working three months later. If there is no response to faradism, there is a 90 per cent chance that the muscle will not be working at three months. This faradic testing shows clearly which muscles are congenitally denervated and gives an indication which ones may possibly recover. A full account of any deformity that is present is also recorded and it is interesting to note that the type of deformity that is present corresponds very closely with the results of faradic stimulation. For instance, if a child has flexed adducted dislocated hips, recurvatum of the knee and calcaneovarus of the foot, we find that the response to stimulation is only present in the hip flexors, abductors, quadriceps and tibialis anterior - that is, innervation from the first four lumbar segments with paralysis and deformity which I have defined and which have been found to correspond when necropsy studies have been done of the affected muscles.

OPERATION

If there is no kyphosis in the lumbar spine, the closure of the spinal lesion is not a difficult matter. It is important that the baby should be operated on in proper pediatric neonatal circumstances on a heating pad. Blood loss is best estimated by using a water bath into which the sponges and drapes are put and estimation made to within 1 cc. Blood is replaced as needed if the loss is more than 15 to 20 millilitres. The area is cleaned very carefully to avoid any additional trauma.

The entire neural plaque is preserved. The surrounding membrane is removed. The dura is carefully dissected up and we nearly always find that is is not too difficult to close the dura over the nervous tissue to make it watertight. Nerve roots are very carefully preserved. The skin is mobilized around the side to the umbilicus and in most instances closure is reasonably feasible though there may have to be a relaxing incision. It is important to relieve tension on the wound by application of a suitable suspensory corset in which the baby is placed in the incubator. We have usually avoided the performance of extensive flaps which seem to break down more easily than a carefully made vertical closure.

A particular problem occurs in children with severe kyphotic deformity of the spine. In these, closure of the skin is almost impossible. The spinal cord is stretched over a bony hump and break down of the skin is almost inevitable. To deal with this difficult situation, we have devised osteotomy resection of the spine with removal of one and a half or two lumbar vertebrae, the vertebrae being taken out from between the spinal nerve roots after carefully dissecting the aorta away from the front of the bodies. The vertebrae can be held together by fine pins and strong silk suture. Another lesion that is unusual is one which presents with a hemimyelocele. In these patients, there is unilateral paralysis and the plaque is only associated with one half of the spinal cord, the other half passing down in its own dural tube. These lesions are always associated with a diastematomyelia. We believe that the central bony process should be removed as well. We do not use systemic antiobiotics but we sometimes use local antibiotics.

MANAGEMENT OF THE CHILD WITH SPINA BIFIDA

Alfred Defalco, M.D.

The primary aim of the urologist should be the preservation of function and structure of the urinary tract, which in children with spina bifida is so liable to damage. With this as a primary aim it is essential that a diligent and aggressive approach be applied to this problem, for the natural history of the disease does not allow for any equivoation or oscillation in management. Urologic management in children with this disorder is actually a continuum of events which may begin immediately after birth and continue into puberty and beyond. It is necessary therefore, to follow a course of continuing management for the total life of the child afflicted with this disorder. (Figure 1)

COURSE OF MANAGEMENT

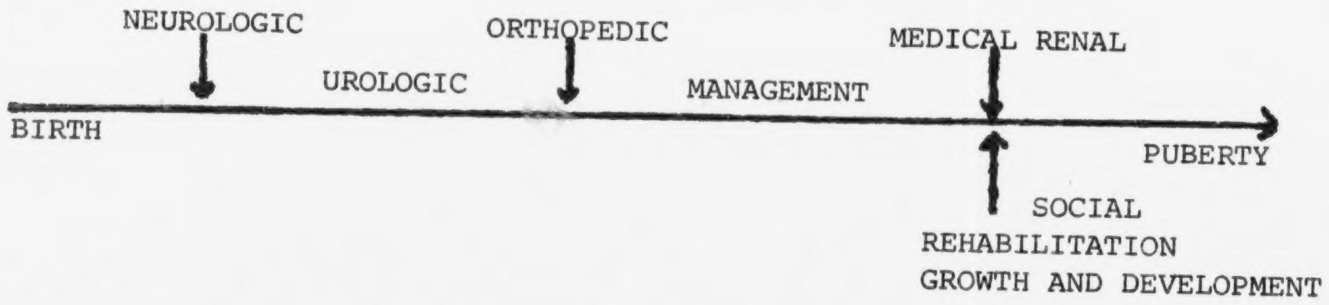

Figure 1.

Shortly after birth of the child with spina bifida, the urologist is primarily concerned with establishing the structural and functional baselines upon which decisions are founded. It is understood that neurologic or neurosurgical problems immediately after birth are the more demanding and certainly supercede any others. Shortly thereafter other physicians are intimately involved with establishing the earliest possible program of rehabilitation, with preservation of muscle groups upon which later motor functioning relies. When the baselines of structure and function relating to the urinary tract are established the children must be watched very carefully for deterioration in this system. Progressive changes in the medical-renal or structural aspects of the patient's urinary tract become more and more important as time goes on. The critical period in management is from one to two years of age. However, it becomes apparent that even after this stage is passed social rehabilitation and growth and development factors assume greater consequence as the child approaches puberty. If early urinary diversion has been performed then as the child enters his adolescent growth spurt the problems of managing his appliances and of integrating his rehabilitation program with his urologic management become important. If diversion has not been performed prior to puberty, the social aspects of urinary incontinence and the child's moving out into society more actively bear heavily on what is done urologically.

If rational judgements are to be made concerning the management of these children the natural history of the condition as it pertains to the urinary tract must be known. It is interesting to follow large groups of children not only for one to two years, but up to five and seven years of age and see what happens to them. (Figure 2)

COMPLICATIONS AT 7 YEARS

REFLUX	30%
STRUCTURAL DETERIORATION	50%
INFECTION	80%
INCONTINENCE	90%
UREMIC DEATH	40%

Figure 2.

By five or six years of age almost 30% have reflux, and by the seventh to the tenth year, fully 50% of these children will have some structural deterioration in their urinary tracts due to the ravages of chronic infection and reflux. The presence of vesico-ureteric reflux in and of itself is not of great import except we know this lays the groundwork for structural deterioration when infection supervenes. It has also been found that fully 80% will have clinically significant recurrent urinary tract infection by the time they are seven. Urinary incontinence can almost be guaranteed in 90% of these youngsters; this influences what is done in the early years, but even more as the children grow to puberty. Finally it is important to note that if children with spina bifida are allowed to be neglected as a group that 40% to 50% will be dead from renal failure by the time they are 14 years of age.

With these discouraging statistics in mind, what can be done to prevent what appears to be a progressive downhill course for these children? First, management must begin shortly after birth with establishment of certain baseline functional and structural parameters. (Figure 3) Ideally, all children with this disease should have an intravenous pyelogram within two to three weeks of birth and preferably yearly thereafter. Cystometric studies to determine basic bladder tone and studies to determine the patient's ability or inability to develop intravesical pressure should be performed just after birth and every one to two years thereafter. Cystourethrogram, a radiologic study to delineate the bladder neck and urethra during periods of either voluntary reflex or forced expressive voiding should be performed shortly after birth and every six to twelve months thereafter. This study gives us good information concerning the general configuration of the bladder, its ability to empty and the presence or absence of relative obstruction at the bladder neck itself or in the area of the external sphincter. Another important parameter to establish early is that of renal function. These studies need not be elaborate, but they should be done every six months. Following the BUN and creatinine with yearly creatinine clearances appears to be adequate to reliably follow the function of the kidneys. Finally, of great importance, are the bacteriologic studies of the urine which should be carried out every three months, beginning at birth and continuing until decisions are made with regard to diversion, and then yearly thereafter.

TEST PROCEDURES

1. I.V.P. - BIRTH AND YEARLY
2. CYSTOMETRICS - NEONATE AND 2 YEARS
3. CYSTOURETHROGRAM - BIRTH AND EVERY 6 MONTHS
4. RENAL FUNCTION STUDIES - EVERY 6 MONTHS
5. CULTURES - EVERY 3 MONTHS

Figure 3.

Management of the urologic problems in the child with spina bifida may proceed along a conservative course for indefinite periods of time. (Figure 4)

CONSERVATIVE MANAGEMENT

IF:

NO DETERIORATION

CYSTOMETRICS - NORMOTONIC, SOME FLACCID

BLADDER NECK - SOME TONE, REFLEX OPENING

MALE

THEN:

MANUAL EXPRESSIVE VOIDING (CREDÉ)

TIMED VOIDING

BOWEL CONTROL

T.U.R.B.N.*

PUDENDAL ABLATION

ANTIBIOTICS

FOLEY CATHETER

EXTERNAL DEVICE

Figure 4.

If in the course of observing the child there has been no deterioration in the upper urinary tracts, if the cystometrics have demonstrated either a normotonic bladder or a flaccid bladder which can be emptied by manual compression, if the bladder neck itself shows some degree of tone or reflex opening on spontaneous voiding, and if the child is a male, a conservative course may be followed. This entails: 1) manual expressive voiding on a carefully timed basis emptying at least every two hours, 2) careful bowel control, and 3) transurethral resection of the bladder neck and pudendal nerve ablation to reduce minor degrees of obstruction at the bladder neck or at the external sphincter. These management modalities are usually supplemented by appropriately applied antibiotic coverage and for males, with the fitting of a satisfactory external collecting device. On rare occasions certain female children can be managed with Foley catheterization and minor antibiotic coverage with very good results regarding continence and preservation of structure and function of the upper tract.

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*Transurethral resection of the bladder neck

The parameters which indicate adopting a surgical approach (Figure 5) are: deterioration of the upper tract, infection which is either persistent or recurrent despite good bacteriologic coverage and a very small hypertonic bladder with no reserve capacity. For most females, there is a tendency to favor early diversion.

SURGICAL MANAGEMENT

IF:

DETERIORATION

INFECTION

NON-RESERVOIR BLADDER

HYPERTONIC BLADDER

FEMALE

THEN:

SPHINCTEROTOMY

VESICOSTOMY

ILEAL CONDUIT

Figure 5.

We prefer to use the ureteroileocutaneous conduit because it appears to be the safest and most appropriately applied surgical procedure for this group of patients. Vesicostomy is used where there is opportunity for intimate association with its technical and management aspects. However, vesicostomy appears to offer very little in the way of advantage over the ileal conduit and the experience of many people throughout the country indicate that it tends to perpetuate infection and is frequently associated with a mumber of stoma problems.

In summary, the management problem may be considered in two temporal periods, the first from birth to 2 years and the second at 2 years of age. (Figure 6)

SUMMARY

BIRTH TO 2 YEARS

I.V.P. AT BIRTH AND EVERY 6 MONTHS

CYSTOMETRICS

CYSTOURETHROGRAM

CULTURES EVERY 3 MONTHS

EXPRESSION VOIDING

BOWEL AND SKIN CARE

T.U.R.B.N.

PUDENDAL ABLATION

AT 2 YEARS

FEMALE INCONTINENCE - DIVERSION

MALE INCONTINENCE - PENILE DEVICE

DIVERSION IF: PERSISTENT RECURRING INFECTION - 3 MONTHS
PROGRESSIVE DILATATION
REDUCTION IN RENAL FUNCTION

Figure 6.

In the time from birth to two years concern centers about establishing baselines as defined by the intravenous pyelogram, cystometrics and cystourethrogram and bacteriologic studies. These should be repeated frequently enough that the urologist is aware of any changes which might be occurring in the urinary tract. During this period manual expression may be employed, voiding on a carefully timed basis; attention is also directed to bowel and skin care, which can be a difficult and tedious problem. If there is some evidence of bladder obstruction or external sphincter spasm, one might consider a transurethral resection of the bladder neck and/or some ablation procedure on the pudendal nerves so as to effect a partial or complete paralysis of the external sphincter.

At the end of two years, a critical point in time appears; for it is at this age that the decision regarding urinary diversion should be made. Achieving urinary diversion at two years of age is not an absolute rule, nevertheless early urinary diversion is going to prevent later damage to the urinary tract in many children. In addition, both patient and parental acceptance of the procedure is much better, and by the time the child is going to school and becoming more socially active the problems with managing the device are greatly reduced. If there is persistent incontinence in older female patients, these are immediately recommended for diversion. However, if the female child is maintaining periods of two to three hours per day continence or urine with the ability to express her bladder by manual method, if her urine is sterile, and if her urinary tracts have not deteriorated, there is a tendency to defer surgery and to observe the child in ensuing years. Fully half of this latter group of children develop some type of persistent or recurrent troublesome incontinence, and by the time they are adolescent we find that many of them are best handled by urinary diversion.

In males, at two years, if there are signs of persistent or recurrent infection, if there is progressive dilatation or destruction of the upper tracts and/or reduction in renal function, then these males should immediately be diverted. It is interesting that many males will come to this point with perfectly normal upper urinary tracts and persistent incontinence which can be handled very easily with a penile device. These children are frequently managed conservatively as are the girls who have periods of dryness effected by manual voiding. Many of the male children who wear penile devices and have perfectly normal upper tracts and no infection will frequently attain adolescence doing quite well, but a significant number of these also have enough trouble with incontinence to warrant urinary diversion for hygienic reasons if for no other.

It is evident that the last chapter has not been written on the management of children with this disorder. A great deal of research activity is contemplated for the future and if properly executed can add additional useful years to the lives of these children as well as guarantee structural and functional preservation in their urinary tracts. (Figure 7). The primary research interest centers about the area of electronic controls.

RESEARCH EFFORTS

ELECTRONIC CONTROLS

COLLECTION DEVICES

URINARY DIVERSION

INFECTION CONTROL

PARAMETERS OF TRAINABILITY

Figure 7.

Basically the question here is whether electronic controls will effect urinary incontinence by means of external sphincter or bladder neck stimulation. If this could be done the child could void at will by either inducing or breaking an electronic stimulus from an implanted smooth muscle pacemaker. Another area of vital interest is the perfection of more appropriate collection devices both for male urinary incontinence and for those children who have ileal stomas. Infection control is still of vital importance and there is an appropriate research interest in trying to control the sources of bacteria which eventually invade the urinary tract. In this area we are working actively on trying to find effective means of either keeping the bowel sterilized or empty enough so that bacteria will not be spread from this source as actively. We are also interested in the possible use of hormone agents such as estrogen to condition the lower urinary tract in order to increase its resistance to infection.

Finally, one of the most interesting areas of research activity is establishing those parameters which will allow us to predict which children will be trainable with passage of time. In this area we are trying to find out whether or not certain factors such as the detrussor tone, bladder emptying dynamics and flow rate, perineal floor activity, and reflex activity of the urinary sphincters have any predictive value with regard to the trainability of the child at some later time. I think we are already convinced that if a child has relatively good detrussor tone and some activity of his perineal fllor and can indeed develop an intravesical pressure either by reflex or external crede that he may be in the small group of children, (probably only 5%) who are trainable and will go through life with a normal upper and lower urinary tract.

In summary, urologic management of children with spina bifida and meningomyelocele is a long-term process which begins at birth and extends through puberty into early adult life. It requires diligence and early decision-making at several critical points in time, all of which should be done on the basis of factual material collected over the child's first one to three years of life. We have tended to limit our major decision-making to the second or third year of life and are more inclined now toward earlier urinary diversion in order to protect the upper urinary tract and to effect a state of urinary continence which will allow the child to enter his social and rehabilitation activities less encumbered. Finally there are several research activities which bear on urologic management of spina bifida. There are some significant contributions to the ultimate aim of preserving function and structure in the urinary tract, thus contributing to a better adjusted person.

TREATMENT OF CHILDHOOD HYDROCEPHALUS WITH A NEW SILICONE-RUBBER SHUNTING DEVICE, " THE DENVER SHUNT ."

A COMPARATIVE STUDY

Wolff M. Kirsch, M.D.*, John B. Newkirk, Ph.D.**
Paul Predecki, Ph.D**, Janet M. Stewart, M.D.*

Pediatricians and neurosurgeons alike are acutely aware of the complications which may attend the placement of mechanical devices for the relief of hydrocephalus. The problems which may arise after the insertion of a cerebrospinal fluid (CSF) "shunt" are well recognized and have been extensively reviewed (3) (5) (15) (16) (17). For the most part, these problems consist of blocking of the device or its catheters, especially due to accumulation of particulate material, excessive protein contamination of the CSF, or infection. The latter may be associated with the use of non-autoclavable devices with metallic components, or complexly recessed plastic devices with relatively stagnant cul-de-sacs. Both of these structual features favor protein desposition in a foreign body - an excellent nidus for eventual bacterial colonization (6) (14). Despite these drawbacks, the insertion of ventriculo-jugular or ventriculo-peritoneal CSF "shunts" has proven to be a worthwhile and reasonably effective procedure for the control of hydrocephalus. (4) (9). Currently available shunting systems are expensive, and this fact limits the availability of the device and its insertion to a restricted population of patients with a favored social setting.

Our objective has been to develop an inexpensive device of simple, yet rugged construction, devoid of metallic parts, with flow characteristics allowing the passage of fluid of high viscosity or with particulate material contamination. This report deals with the use of a new system ("The Denver Shunt"), which may meet the above specifications, in 30 pediatric cases at the University of Colorado Medical Center over an 18 month period. This clinical experience will be compared to our experiences with other shunting systems over an approximately equivalent time span.

*Division of Neurosurgery, Department of Surgery and Department of Pediatrics, University of Colorado Medical Center, Denver, Colorado
**Denver Research Institute, Denver University, Department of Metallurgy, Denver, Colorado

The Denver Shunt is available from Biomaterials, Inc., 2551 E. Floyd Avenue, Denver, Colorado

Description and Flow Characteristics of the Denver Shunt

The shunt bodies are molded and vulcanized in one piece from "silastic"*. The proximal valve of the shunt is a simple slit contained within the molded part. The distal valve consists of two slits at the exit end of the distal tube (Figure 1). The simplicity of design runs counter to the current developmental trend of more complicated and expensive shunting systems (1) (10). Both the proximal and distal slits in the shunt act as one-way valves. A positive pressure gradient in the direction of flow causes the valves to open permitting gradient (Figure 2). A negative pressure gradient forces the valve lips closed and prevents back flow. The net effect is a rectification in proportion to their intensity.

Bench tests of the Denver Shunt, performed with solutions of widely varying protein concentrations including the addition of whole blood to artificial spinal fluid, show favorable lack of sensitivity of the flow rate through the shunt with respect to protein concentration (Figure 2). It would appear that the increasing viscosity of the protein solution is balanced by some counter effect such as an increased adsorption of protein on the slit lips (11). This in turn causes the slit corners to be pushed further apart and dilates the slit to balance the viscosity increase. The marked facilitation of the flow rate of distilled water observed during _in vitro_ testing upon the addition of minute amounts of albumin. If distilled water is passed through the shunt, and the slit surfaces are clean, the surfaces tend to knit and stop the flow even at 10 cm. pressure. This tendency has not been observed with protein solutions above 2-3 mg.%, and in fact, there is a slight but definite facilitation of flow at high protein concentrations.

Technical details regarding the mechanics of the device and methods of implantation have been described in a previous publication (8). A principal advantage of the shunt appears to be that of providing a simple pumping action through a proximal slit valve capable of passing significant amounts of particulate matter without clogging. Should the valve become clogged, the absence of rigid parts may permit clearing of the system by simple digital distortion of the proximal valve. This maneuver has worked clinically on several occasions, but has failed in two cases where plugging occurred at the distal slit. The latter malfunction may necessitate revision distally with elimination or relocation of the distal tubing. Since the valve is constructed entirely of silicone-rubber, tissue compatability is maximized and thrombogenicity is minimized. The shunt may be autoclaved and stored dry.

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*Medical grade Silicone rubber manufactured by Dow Corning Co.

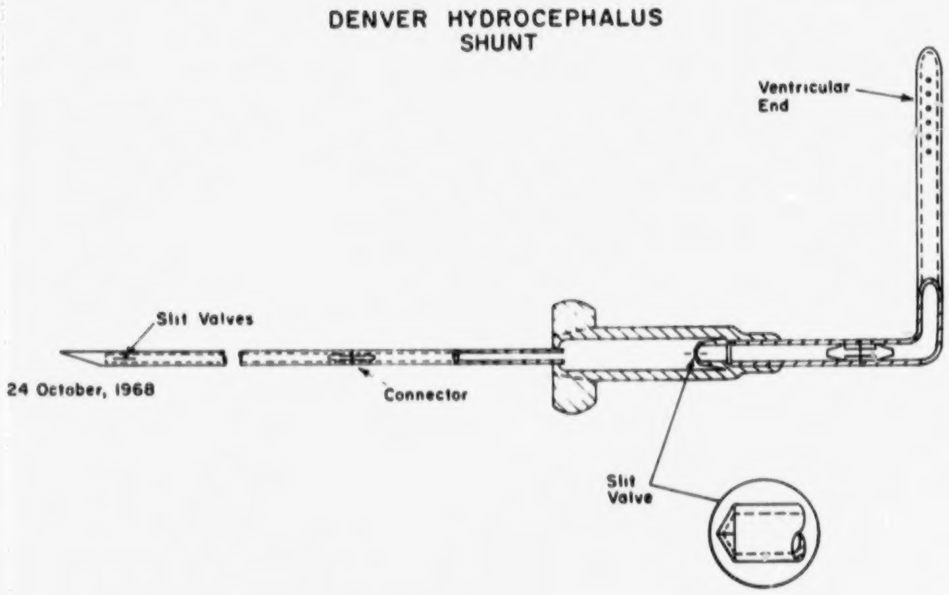
DENVER HYDROCEPHALUS SHUNT
Ventricular End
Slit Valves
24 October, 1968
Connector
Slit Valve

Figure 1. Diagram and Photograph of the Denver Hydrocephalus Shunt. Note attitude of proximal slit valve perpendicular to cranial vault.

FLOW CHARACTERISTICS

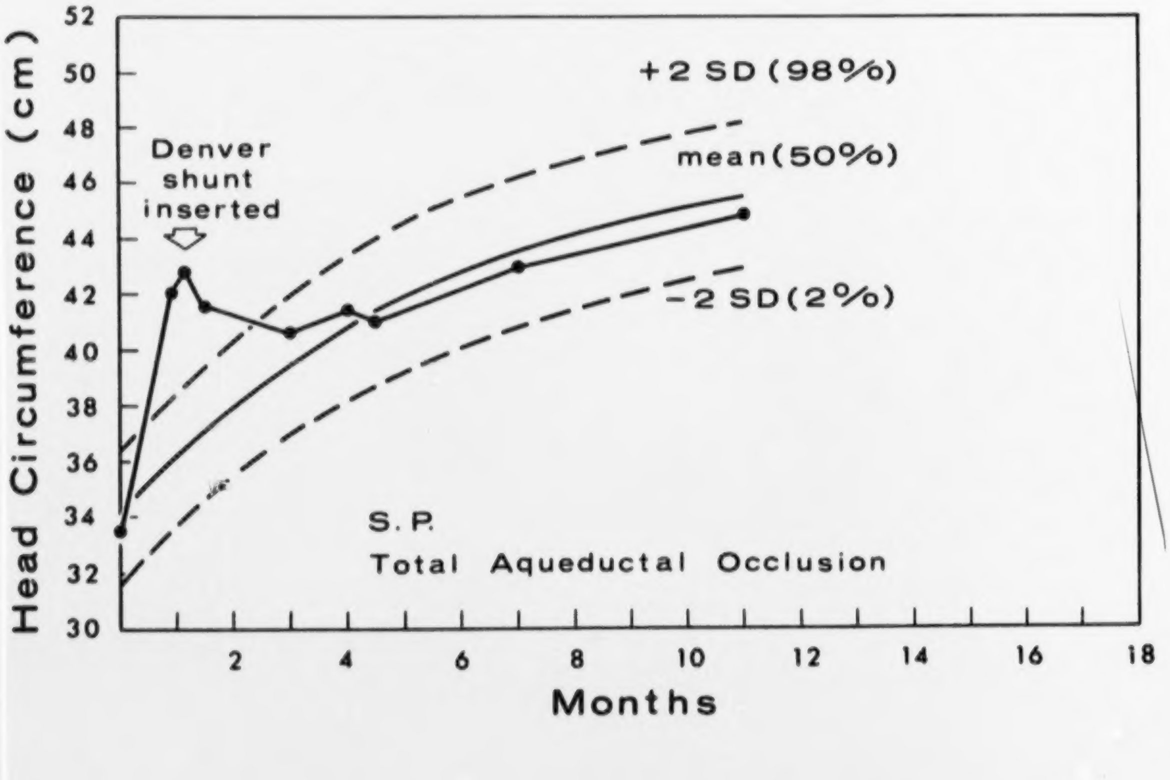

Figure 2. Flow characteristics of a typical "high-flow rate" Denver Shunt. Hemoglobin addition was a 1:200 dilution of fresh blood in artificial spinal fluid containing protein at a level of 1 gm.% Distances denoted signifies 2 standard deviations of the mean at each point (5 separate determinations at each point).

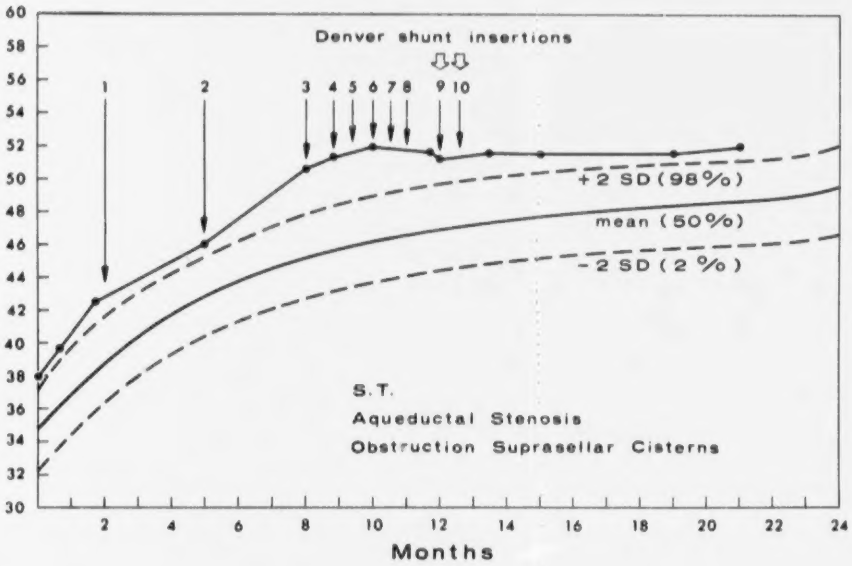

Figure 3. Head circumference progression with chronology of operative procedures, hospitalizations for case 1. (1) Initial insertion of ventriculo-jugular (V-J) shunt and (2) New shunt inserted, (V-J) proximal catheter blocked (3) New shunt inserted, (V-J) distal catheter blocked (4) New shunt inserted, valve mechanism faculty, ventriculo-peritoneal diversion, (V-P) (5) infected (V-P) removed (6) (V-P) shunt reinserted (7) Cellulitis about scalp wound (8) (V-P) shunt removed, valve not functioning (9) Denver Shunt inserted, ventriculo-pleural (10) Ventriculo-pleural diversion converted to ventriculo-innominate.

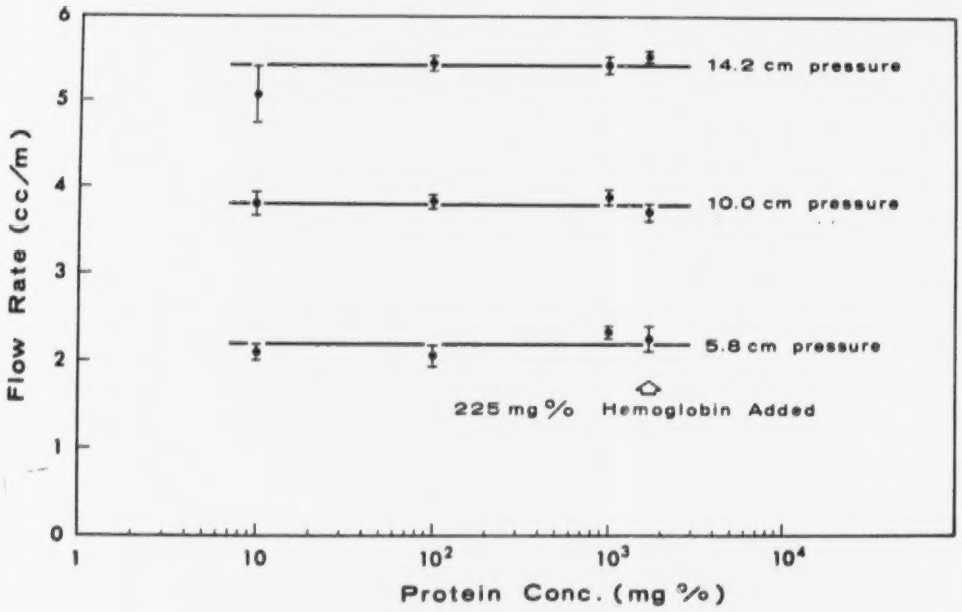

Figure 4. Head circumference progression in Case. No. 2.

Clinical Experience with the Denver Shunt

Prior to clinical application of the device, Denver Shunts were implanted chronically for 3 to 6 months time in dogs (Lumbar subarachnoid-peritoneal diversions). Testing in this manner demonstrated no significant tissue reaction about the silicone tubing as well as patency of the diversion. Denver Shunts were initially used in troublesome cases of childhood hydrocephalus, which had proved refractory to treatment by multiple shunting procedures with other devices. As confidence and experience with the device increased, the system was employed at the initial operative procedure for the management of hydrocephalus, both in children and adults.

The University of Colorado Medical Center series of Denver Shunt insertions in children consists of 30 patients extending over an 18 month period at the time of this writing. Conditions which have been treated and effective types of cerebrospinal fluid diversions are given in Table.1. It is appreciated, however, that examination of tabular compilations concerning the treatment of hydrocephalus, especially in children, tends to give a superficial and incomplete portrayal of the types of problems encountered, overlooking the individual and unique circumstances of many of these cases. In order to convey our experiences in a more meaningful way, and relate problems as well as successes which we have encountered with this shunting device, two representative case histories are reported. Denver Shunts, when utilized will be referred to specifically.

Case No. 1: Aqueductal Stenosis, Arnold-Chiari malformation, and obstruction of the suprasellar cisterns in a child.

This infant boy was first seen by the neurosurgical service at the Colorado General Hospital at the age of 6 weeks in August 1967 because of a progressively enlarging head. The child was otherwise in good health. Ventriculography demonstrated obstructive hydrocephalus with partial aqueductal occlusion and complete obstruction at the level of the suprasellar cisterns. A ventriculo-jugular shunt was inserted but the child's head continued to enlarge (Figure 3). Three months after his first shunt operation the child became irritable, lethargic and began to vomit. He was readmitted to hospital with increased intracranial pressure, bilateral sixth nerve paresis, and a non-functioning shunt. A new ventriculo-jugular shunt was inserted and the patient temporarily improved. His head size continued to increase at a rapid rate, and he was readmitted to hospital for conversion of the ventriculo-jugular shunt to a ventriculo-peritoneal diversion. Within 4 days after discharge he was readmitted again because of lethargy, vomiting and fever. The ventricular fluid and shunt mechanism were found to be infected with E. Coli. The shunt was removed and he improved with systemic and intraventricular methicillin therapy. A left sided ventricular-peritoneal shunt was placed. The child was hospitalized again because of superficial cellulitis over the site of subcutaneous scalp reservoir, which subsided after the application of silver nitrate soaks. The patient's eighth admission was again for signs and symptoms of increased intracranial pressure, and on June 11, 1968 a Denver Shunt (high flow rate) was placed as a ventriculo-pleural diversion (left side). The child required

TABLE I.

Clinical Resume of Pediatric Cases Treated with The Denver Shunt

Children (30 cases)

Conditions Treated	Communicating Hydrocephalus	
	Idiopathic*	10
	Post E. Coli Meningitis	2
	Post Pneumococcal Meningitis	1
	Post Tuberculosis Meningitis	1
	Obstructive Hydrocephalus	
	Aqueductal Stenosis	9
	Arnold-Chiari Malformation	5
	Tumors	
	Glioblastoma of Cerebellum, Pons	1
	Optic Glioma	1
Final Effective CSF Diversion **	Ventriculo-Peritoneal	24
	Ventriculo-Jugular	5
	Ventriculo- Innominate	1

* 3 Patients had grossly hemorrhagic ventricular fluid at time of shunt insertion.

** Final shunting procedure which effectively controlled hydrocephalus. All post-meningitic cases received ventriculo-peritoneal diversions.

readmission two weeks later because of marked dyspnea and irritability. Chest x-rays demonstrated a massive hydrothorax on the left with a shift of the mediastinum to the right. Multiple thoracenteses were required to alleviate respiratory distress. On July 9, 1968 a thoracotomy was performed and the distal end of the Denver Shunt was placed in the left innominate vein. Since this procedure (12 months at the time of this writing) the child has not required rehospitalization, has had no further increase in head circumference, and no evident increase in intracranial pressure.

Case No. 2: Aqueductal Stenosis in a child.

An infant girl was admitted to the Colorado General Hospital at the age of 1 month because of an abnormally large head. The child appeared healthy on admission and no abnormalities were noted on transillumination of the skull. After ventriculography demonstrated hydrocephalus secondary to complete obstruction of the aqueduct, a high flow rate ventriculoperitoneal shunt (Denver) was inserted. Since this procedure the child has done well and her head circumference has remained within normal limits (Figure 4). She is now 18 months of age and her behavior and development are so far considered to be normal.

Comment:

Our clinical experiences with the Denver Shunt have indicated that the device provides safe and effective control of childhood hydrocephalus, and in addition confers several significant advantages over other currently available systems. As the data below indicates, the incidence of serious and potentially lethal complications (ventriculitis) have been significantly reduced at our institution since adoption of the system. In addition, the incidence of recurring hospitalizations for valve revisions has been reduced to one third of our previous requirements with other devices. The need for revision imposes considerable psychological and economic burden upon families who are usually ill-equipped to meet these circumstances. The duration of hospitalizations of children with ventricular fluids containing elevated protein or particulate contaminants (who would ordinarily await shunting until their fluid returned to near normal character), are considerably shortened by the use of a system capable of diverting fluid of any character. The simplicity of the device and insensitivity to the presence of air in the valve itself, eases the technical maneuvers required for insertion. Air in other shunts may significantly alter flow characteristics or block the device.

To compare our Denver Shunt results with those obtained with other systems we have considered any revision of a shunt within the first year of placement in a child as a failure irrespective of cause. Causes for revision of the Denver Shunt have in our experience been different than those responsible for failure in other systems. In Case No. 1 for example, the Denver Shunt required revision because it resulted in massive hydrothorax, necessitating the use of the innominate vein as a recipient site. In fact, seven of the fourteen Denver Shunt revisions in children were necessitated because of failure of cerebrospinal fluid to be effectively resorbed on arrival at the recipient site. Inadequacy of the peritoneal resorbing surface resulted in CSF collecting along a subcutaneous tract in the region of the abdominal wound. Plugs obstructing the distal slit necessitated 3 of the 13 revisions, but in no instances has the proximal valve mechanism or catheter been found to be obstructed. We have so far had no instances of ventriculitis, septicemia, or endocarditis in our pediatric cases, though one case of ventriculitis has been associated with an adult placement. In one infant an abdominal wound became erythematous three days after a ventriculo-peritoneal shunt insertion. The shunt was removed, both the wound and device cultured, but no organisms were identified. The infant was subsequently reshunted (Denver Shunt) without complication. Two Denver Shunts were revised in children because of disconnections at tubing anastomosis distally. In one case a Denver Shunt did erode through the scalp, but no infection resulted. This patient was a small child with an exceptionally large head and attenuated scalp, in which the shunt body was placed too close to the suture line. The device was well tolerated in the remainder of cases.

For purposes of comparison, the frequency of early difficulties with the Denver Shunt has been related to the total duration of shunt implantation by summing the total number of weeks these devices have been in place for all cases. This value is expressed as total "shunt-weeks". As of this writing, the interval time for the Denver Shunt is approximately 680 "shunt-weeks" in 30 children, extending from February, 1968 to July, 1969. This value is to be compared with our experiences in 32 cases incorporating other shunting devices during a period of time extending February, 1967, to March, 1968, totaling about 640 "shunt-weeks" (Table II).

There are numerous factors which might be responsible for shunt dysfunction within the first year of its installation. Attention to technical detail, with due respect to the surgical procedure itself, has been repeatedly emphasized as being the most relevant feature for successful shunting (9). We would certainly concur with this statement. Despite intentions to derive valid comparisons, there is no doubt that bias could have been introduced into this study. Our familiarity with the Denver Shunt may have resulted in an extra measure of surgical care during the process of its implantation. It should be stated. though, that the vast majority of these procedures, irrespective of the shunting device utilized, were performed by our resident staff with a discontinuity of personnel. We have disseminated these devices to other institutions, and preliminary reports so far confirm our conclusions regarding the effectiveness of the device (2) (7) (13). Types of cerebrospinal fluid diversion were not significantly different in the two series. Small infants, or individuals with particulate contamination or high protein levels in the cerebrospinal fluid received ventriculo-peritoneal diversions, whereas other cases have for the most part been treated with ventriculo-jugular insertions.

Uncertainties exist in the categorization of early causes for shunt failure in any series, but certain inescapable conclusions have been made with regard to the Denver Shunt. Our need for shunt revision in children within the first year of placement has been significantly reduced (Table II). Failures of the Denver Shunt have primarily been those difficulties associated with inability of the recipient site to resorb the diverted cerebrospinal fluid, and secondarily, plugs at the distal slit valve. These difficulties are

TABLE II.

Comparative Early Complications of Shunting Procedures for Hydrocephalus
(Denver Shunt and Other Systems)
Children

	Other Devices	Denver Shunt
Cases	32	30
Revisions	38	14
System Blocked	24	3
Proximal catheter	16	0
Shunt	5	0
Distal catheter	3	0
Infections	6	1 (Suspected, no organism identified - See text)
Subdural hematoma	2	0
Failure of recipient site to resorb CSF	0	7
Miscellaneous (tubing disconnection, scalp erosion)	3	3
Deaths	6 (3 deaths attributable to ventriculitis)	1 (Patient with pontine and cerebellar glioblastoma)

apparent soon after the shunt has been inserted. Obstruction proximally or valve failure has not yet been encountered with the Denver Shunt. The incidence of early infectious complications in childhood cases receiving Denver Shunts appears to be significantly reduced. In children treated with other devices there were three deaths directly attributable to shunt infections. Our incidence of early infections using shunts other than the Denver Shunt is comparable to infection rates reported in other series (3) (5) (15) (16) (17).

We have on several occasions been able to clear "Denver Shunts" which pump with difficulty by pressure over the proximal slit. Three valves, placed in infants with grossly hemorrhagic ventricular fluid, have continued to work without revision. In one case, a Denver Shunt remained functional in a child despite a protein concentration of 5000 mg.% in the cerebrospinal fluid. Reviewing the etiology of shunt failures with systems other than the Denver Shunt indicates that disturbances at the proximal catheter or valve mechanism itself were responsible for at least 24 of the 35 revisions. Debris or clot in the valve mechanism, or valves operating at too high pressures were noted in 5 cases. Perhaps the least qualified complication of a "shunt" insertion is infection, or bacterial growth in the valve system, which was detected in 6 patients who received other types of shunts. Obstruction of the distal catheter, subdural hematoma and tubing disconnections accounted for the remainder of the revisions with other devices. We have not yet encountered a subdural hematoma after the insertion of a "Denver Shunt", despite the use of high flow rate devices.

It should be emphasized that the introduction of any foreign body into the brain, thorax, or abdomen represents a significant biological hazard. If in addition this foreign body has the supervening requirement of continuous mechanical competence in the face of changing _in vivo_ conditions development of a perfect system is unlikely and probably impossible. As our experience with the Denver Shunt increases we will almost certainly encounter infections and mechanical failures. We are describing our initial experiences, which have been gratifying, with a device which we feel offers a practical and economically advantageous means for the control of hydrocephalus. The device provides significant advantages over other currently available systems. These advantages are given in terms of comparative clinical studies as well as flow characteristics of the shunt itself, thus fulfulling certain criteria outlined by others in consideration of the vexing clinical problem of hydrocephalus (12).

Summary

A new shunting device for the treatment of hydrocephalus is described, with flow characteristics which remain relatively constant despite large variations in both particulate and protein concentration of the fluid passing through it. The system has been used in 30 children over an 18 month period and the results compared with the use of other devices over a comparable span of time. Mechanical failures and infections appear to be significantly reduced by the use of this relatively simple and economically advantageous device. Failures of the system have consisted primarily of inadequacies of the recipient site to cope with diverted cerebrospinal fluid.

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MOBILITY AND MENINGOMYELOCELE

Harriet E. Gillette, M.D.

To enable the child with spina bifida to change location of the entire body mass or of a small number of its parts is the goal of all who are concerned with the patient with this disability. Those specialists who deal with systems of behavior other than the motor aspects are concerned with movement in society, whether concerned with intellectual abilities or social acceptance dependent upon esthetic considerations. Regardless of the number of body segments involved, mobility in a child with or without spina bifida is necessary for self-care, recreation, education and vocation.

The infant without motor handicap develops effortlessly into the child who can run with the wind as it raises his kite, then into the professional athlete, or to the typist with intricate finger movements. Mobility in the child with spina bifida may mean functional activity of only one body segment to accomplish a purpose, such as feeding. Or it may mean progression of the whole body through space by one of several varieties of locomotion: crawling, rolling on the floor, propelling a wheelchair, supported ambulation, independent ambulation using assistive devices, or unassisted ambulation.

There are few conditions which present the heterogeneous assortment of varieties of motor behavior seen in spina bifida. The spinal cord lesion may produce:

Flaccidity due to lower motor neuron lesion, a result of anterior horn cell or peripheral nerve impairment
Spasticity due to upper motor neuron lesion, result of interruption of decending tracts
Sensory disturbance
Incoordination, a lack of integration of spinal reflexes

Cerebral dysfunction, a common associated handicap, results in:

Varying degrees of spasticity and/or dyskinesia and ataxia
Perceptual impairments
Intellectual impairments

Societal influences often produce:

Obesity
Contractures from continual faulty positioning
Weakness and atrophy of disuse
Deprivation phenomena due to lack of social, sensory, motor experience

Motion in the normal depends upon many factors, each of which must be present in optimal amount to achieve integration with all the other elements. The more important components of normal movement include:

Sensation: the essential component of movement, for without stimulus there is no response

Perception: the interpretation of stimuli, determines the effectiveness of motor ability, adaption to the environment, deployment of body parts. Though intangible, this function determines the level of motor function within the individual's competence.

Tone: Balanced between flexors and extensors, responding reflexly to stimulation of tone-controlling receptors.

Range of Motion: Unrestricted movement at each joint, consistent with form and function of joints and surrounding soft tissues. Varieties according to body type, age and occupation produce a wide range of normals.

Coordination: A melding of sterotyped pattern movements produces a fluid, effortless, graceful motion which is an indication of a fully myelinated, intact central nervous system.

Postural reflex responses: Range from the primitive reflex shifts of tone in response to stimulation of certain receptor organs, to complex adaptive posturing. Tonic labyrinthine, neck and lumbar responses constitute the means whereby the body adapts itself to space.

Balanced strength: Excessive strength of a muscle group disporportionate to strength of its antagonist is inconsistent with function. The intricate mechanism for joint stability insures an appropriate balance of muscle power about a joint.

Against these requisites for normal motion, contrast their level of functioning in the child with meningomyelocele.

Impaired Sensation: absent or impaired in legs, and lower trunk, there is no stimulus to warn of harmful pressure or temperature, or to signal position or to initiate a motor act.

Distorted perception: disability in this sphere constitutes a serious impediment to mobility, for interpretation of sensation is requisite for its effective use. Lack of knowledge of spatial relationships of body parts, of sequence of acts often prevents use of crutches, appropriate foot placement, stabilization of trunk, and lack of hand skills even though the neuromuscular unit is intact.

Abnormal amount, quality and distribution of tone is usually below normal, with instability of joints. Hypertonia may be present, due to either interruption of pyramidal tracts in their course downward through the spinal cord, or to cerebral dysfunction which may produce the findings of cerebral palsy, with motor behavior of spasticity or dyskinesia.

Restricted ranges of motion: the mechanical problems imposed upon disorders of neuromotor function compound the problem many-fold. Because they are obvious and often amenable to correction by either conservative or surgical means, the sensory impairment is apt to be overlooked and the operator blamed for lack of success of the surgery or the therapy. Hypermobility of joints poses as much of a problem as restriction, and is probably more difficult to manage.

Incoordination: Lack of blending of pattern movements constitutes an impairment of fine skilled acts though not necessarily loss of function for gross motor performance.

Delayed development of postural reflex responses: impairment of both spinal and cerebral reflex movements may prevent the most rudimentary adaption to space, with lack of tonic lumbar responses, crossed extension, placing or stepping, etc.

Unbalanced strength: disproportionate power in muscle groups surrounding a joint results from disturbances of tone, lack of reflex movements, immobility due to perceptual disturbance and lack of environmental stimulation. It is both cause and result of deformity and the point to which treatment is directed.

ENCOURAGING MOBILITY

In the light of what appear to be insurmountable obstacles, what should be done to salvage all possible components of motion to render the child with meningomyelocele mobile?

"Mobility" carries different connotations in different situations and at different ages, all related to the displacement of body mass through space, but all directed toward the final goal of enabling a mature individual to progress from point A to point B to perform a meaningful activity.

The process begins in the nursery, as soon as vital signs are stable. The prone position necessitated by the dysraphia, with or without surgery, is fortunately the desirable one. Legs should be maintained in extension to accomplish the first objective: prevention of contractures. In this period, other contractures may be anticipated in abductors and foot dorsiflexors. It is here that deprivation begins unless measures are taken to prevent it, for if there is no movement of body parts, whatever sensation there is, is suppressed through lack of stimulation.

An inviolate rule of the nursery should be that of performance of ranges of motion to all extremities at least six times a day, with positioning to maintain body segments in correct alignment. Parents should be thoroughly instructed in these procedures and in the reasons for them, before taking their baby home.

As the infant matures, reflex motor responses and sensory and perceptual awareness should be specifically encouraged through activities of rolling, crawling, creeping, knee standing, sitting. Ranges of motion must continue, as contracture formation is a continuing threat. It is appropriate now to provide a means of locomotion, enabling the child to propel himself on such devices as a low stool on casters, or a wheelchair.

Sensory and perceptual development must receive the attention afforded these functions in the normal child through usual play activities; the handicapped child, however, usually receives this attention only through a conscious effort. It has been pointed out that these abilities are a necessity for motor performance.

As postural reflexes develop, they should be utilized in the appropriate position and manner. This will usually necessitate braces and crutches. Support should be adequate to maintain skeletal alignment, but not so great that function is compromised. Bracing elements should be removed progressively as function develops.

A wheelchair is a necessary tool as well as a convenience for many children and adults. It provides mobility which might never be attained by ambulation. In many instances, a wheelchair will be the only means of mobility, and it is both useless and frustrating to attempt to achieve an unrealistic goal of ambulation if necessary elements for it are not present.

Success of the effort to provide mobilization for the child with meningomyelocele is dependent upon the accuracy of determination of proper goals. These are changed with changing state of maturity, but basic requirements remain the same for whatever kind of mobility is appropriate to the individual's capacity:

Complete use of sensory capacity
Full range of motion of all body segments
Use of all postural reflexes
Provision of appropriate means for locomotion

PROBLEM SOLVING IN MANAGEMENT

Margaret L. Hill, R.N., P.H.N., M.N.

The child with spina bifida, as you have found in the course of your work, presents problems of tremendous magnitude to himself, to his family and to the community. No other group of patients offers a more challenging array of remediable complications,whether natural, acquired, infectious, developmental, metabolic and surgical. The challenge should not be to rehabilitate, but to maintain as near a normal developmental pattern as possible.

The recent trend has been to establish unified comprehensive care programs for children with handicaps. Unfortunately, there are still centers in which the child's care is fragmented among specialists. Parents become frustrated as no one person is responsible to meet these life long handicaps. In addition, there frequently is no follow-up of the recommendations when the child returns to his own community.

The multiple defects of a child born with a meningomyelocele concern many specialists. Therefore, a well conceived program of continuity of care is essential. These children are the classic example of the necessity of collaboration and cooperation in a well coordinated team. Initially the neurosurgeon repairs the back and may or may not have to shunt to control the hydrocephalus; the urologist is concerned with preserving the kidneys; the orthopedist maintains function in the feet, legs and hips by casting, surgery, splinting, positioning, and with the physiatrist prescribe the functioning and ambulation program. The social worker evaluates the family dynamics and provides counseling; the psychologists are concerned with the developmental assessment; the nurses provide the nursing care and are responsible for coordinating patient care between hospital, clinic and home. The public health nurse, family physician, and/or any other discipline in the community that are involved with his care are also important members of the health team; however all these services cannot function in a disconnected fashion. To coordinate the diagnostic and treatment services for these children, the pediatrician who is in charge of the continuum of care usually directs the program. The team members along with the program director confer on their findings and together approve a therapy plan. A network of communication is set between the center,child's physician, public health nurse and/or anyone else in the community that is involved with providing continued care.

Family dynamics: Before we can discuss management, or the "how" of problem-solving, it would be helpful to look at some of the observain family dynamics that we have seen, for an understanding of them is essential in planning a realistic treatment program for the child and his family.

We need to consider the child as a child. We are so intent upon the multiple disease processes, that we often need to be reminded that our patients are children who have needs, reactions and problems common to all children. Appropriate for the age level, they need mothering, environmental stimulation, discipline, socialization, well child supervision and immunization. Parents are the other half of the picture as they are an inseparable part of the child's environment and his development. They, too, have problems that are common to all parents such as: role confusion, feeling guilty when their child has problems such as enuresis, thumb sucking, aggressive behavior, etc.

In addition to everyday problems there are those which the parents have to live with over a span of time. One of our mothers, when asked what was the hardest part of her daughter's care replied, "I can take care of the day by day problems, but it is just knowing that it will go on and on and on -- never ending, is what gets to me!" They have a child who is chronically ill. One problem may be solved and another crops up. If the child has a shunt, there is a chronic anxiety caused by fear of shunt failure. There are repeated hospitalizations. If mother stays with the hospitalized child the siblings may be neglected. The financial drain is considerable even when the family has medical insurance. The mass of work which must be done by a conscientious parent is incredible, with constant medication, bracing, lifting, multiple trips to the doctors and physical therapists. It is surprising how many in-laws try to defend their own ego, implying that the defect is hereditary but probably not from their side of the family. These are but a few of the problems confronting parents.

What is the impact of all those problems upon the family? Many of our mothers are chronically depressed. Some of the parents withdraw from the community, friends and neighbors. They feel self-conscious about the reaction of other people to their handicapped child. Some parents displace their anger on the sibling because it isn't "nice" to be angry at a handicapped child. Parental anger, guilt and rejection are often manifested in patterns of over-indulgence, over-protectiveness, inability to set limits, and inability to help the child develop self-help skills. There are parents who sabotage the treatment by not following through on recommendations which would enable the child to develop independence.

What happens to the children who have never been allowed to develop independence? They develop an apathetic, dependent, passive behavior pattern. They become masters at manipulating their environment, getting others to perform for them, bringing their environment to them. They never learn to progress in their development of independence from receiving to giving. Parents, particularly mothers, become frustrated because they are chained to a dependent child. As the child reaches adolescence, parents become concerned about what will happen to the child when they are gone.

The father also has feelings and needs help in facing crisis, although he often won't admit it. In our culture, man has been brought up to feel ashamed to show his feelings. As a result of this social stigma, the father feels excluded from participating in the care. He doesn't know how to become involved, and is unable to swallow his pride. The public health nurse is in a key position to help him become involved by arranging a home visit to fit into his work schedule. There may be some simple device that the child needs such as a scoot-board for locomotion that Daddy can make. This indirect approach in getting Father involved works in the majority of cases.

The siblings are often orphans of the treatment system. Mother is so involved in the care of the handicapped child that she hasn't time to spend with them, or thinks she hasn't. As a result, behavior problems such as enuresis, underachieving in school and aggressive behavior will appear. This type of behavior usually calls for counseling. The public health nurse or school nurse is in a key position to assess this type of behavior and refer the child and his family for future follow-up. How siblings accept their handicapped sibling is usually dependent upon the parent's attitude. If he is treated like a regular child except for his defect, children will tend to accept him. Brothers and sisters need to see conformity, politeness, generosity, and helpfulness. This type of behavior is even more important for the handicapped child as this convinces him that he is a regular member of the family.

The How of Management: An attempt to understand parental anxieties will assist in the management of the emotional complications which are a part of the child's total care. Feedback from the public health nurse regarding the patient's progress and what is really happening in the family will assist the health team in identifying areas of stress or strengths which can be incorporated into the total treatment program. Realistic goals need to be established for the family and patient. Flexibility in the program is critical to meet the ever changing needs of the child and his family.

How can we assist the child and his family to optimal development? In conjunction with their medical management, an important goal for these patients is to get them upright and mobile by use of adaptive devices as close to the normal age as possible. Their emotional development, hand-eye coordination, spatial perception, development of head and trunk control, upper extremity strengthening, and their general concept of the world about them are all dependent to some extent upon the stimulus provided by being upright and mobile. It is important that the entire health team be unanimous on the ultimate goal for the patient so that parents do not become frustrated by differing answers, or by unfounded hopes for the patient's future function. Pushing him toward a level of function for which he is physically incapable produces not only a high level of frustration, but emotional and personality problems as well.

A program of splinting and positioning to prevent the development of postural deformities can be begun in the neonatal period. As the child progresses, adaptive devices for positioning and mobility are introduced. Being held by the mother or nurse provides the first experience in mothering in the upright or semi-upright position. Tactile stimulation can be provided by the ward nurse by touching the infant or rubbing his cheek as she goes by the crib. The crib can be turned so he can become a part of the world about him. The infant tilt seat provides a means for getting the infant up and a part of his environment. This allows him to be with mother when she is working. He not only hears her, but can see what is going on about him. If the meningomyelocele has not been excised, a sponge pad can be placed in the infa-seat around the lesion.

As the baby reaches the sitting stage, he should be placed in a high chair or baby tender. If these are not available, a sitting device with support straps may be used; this is easy to make and will fit on any chair. A brightly colored toy such as plastic teething beads may be placed on the tray in front of him. Anchoring the toy to the tray's edge with a string saves the mother pick-up time. This allows the infant to develop not only hand-eye coordination, but also sitting balance which is often latent in these children.

As the child progresses to the crawling stage, but is unable to crawl, a scoot board will provide him with a means of locomotion by which he can investigate his environment. This also develops the shoulder girdle and upper extremity muscles for later crutch walking and/or transfers.

A small nail barrel is an excellent device to facilitate standing balance with or without braces. By anchoring the barrel on a platform with coasters, it can be moved from place to place. Children like this gadget as they have freedom of movement in it. A heavy plastic waste basket or garbage can also serves the purpose. The child may also be strapped on to a tile board for standing even without braces. Weight bearing is essential in decreasing the number of pathological fractures, and in promoting bone healing and strength following surgery.

Early ambulation, between one and two years of age, in a tiny tot wheelchair or any adaptive device provides the child with a means of investigating his environment,an essential aspect in developing independence. Very young children learn to navigate the chair and to transfer without being taught.

Corrective orthopedic procedures are being attempted at an earlier age so that the child can become ambulatory at the appropriate age level; however, the importance of continued proper positioning and good body alignment in the prevention of deformities cannot be underestimated at any time. Proper positioning can be demonstrated and discussed with the patient and family, but the public health nurse in the community sees what the patient does or doesn't do at home. Many times the only contact with the patient and his family between clinic visits is through the nurse. Periodic progress reports from her are very important and even more so if she has observed a change such as increasing contractures and deformities, or ill-fitting braces. The nurse's report allows the team to act promptly instead of waiting until the patient comes to the clinic for his routine visit.

Since many public health nurses are responsible for the home therapy program when there is no registered physical therapist available, she needs to be knowledgeable in putting on braces, checking proper fit to prevent pressure sores, teaching care of braces, and maintaining proper crutch height for good posture, beside performing therapy. Eventually , it would be anticipated that the parents can carry on with the program under the nurse's supervision.

Because of apathy and dependent behavior that has been fostered, many children do not perform these activities of which they are capable such as dressing, toileting, personal hygiene, feeding , locomotion, and socially appropriate responses. Our basic concern is to help the handicapped child fit into society by one means or another. For a selected group of patients, behavior modification may alter the child's behavior by rewarding desirable performance. The community nurse has a primary role in insuring the success of this type of program as she can be an effective behavioral therapist in the home.

Play therapy may also be a very important part of a behavior modification program. Studies have shown that the child with a handicap has fewer toys, books and less opportunities for socialization outside the home than his siblings, probably because parents are so concerned with the child's problems that his basic needs as a child are often overlooked. Toys and play materials give the child not only opportunities for emotional release and incentives for the development of motor skills but are also a means of socialization.

Crafts of a creative nature are essential for these children. Play activities that increase perceptual and conceptual function should be encouraged; examples are puzzles, tinker toys, building materials.

Whenever possible outdoor activities should be encouraged. Swings, slides and sand boxes are useful equipment. Play that will increase muscle power in the trunk and upper extremities should be encouraged (i.e. ball handling, clay, pounding boards, chinning bars etc.) The child's social development is furthered whenever he can participate with his family or peer group in playing games. Cooking can be used as a form of play for both boys and girls.

The importance of play in the development of children is receiving increasing emphasis. The child learns in steps -- looking, reaching, grasping, and finally exploring. To develop these tasks, he needs visual, sensory and auditory stimuli. Because all children develop at different rates, toy selections should be based on developmental steps rather than age level. They do not have to be elaborate; how they are used is most important.

The natural tendency of mothers is to over-indulge the child with food. Overeating and inactivity combined with a limited caloric need as compared to the normal child leads to obesity. Emphasis should be placed therefore on maintaining a below normal weight/height ratio, for minimum weight not only assists mobility but decreases pressure sore formation.

Achievement of bladder and bowel control is an integral part of achieving self-identity and self-conceptualization. The child with meningomyelocele rarely achieves this control because of the deficient innervation of bowel and bladder.

Of all the problems to which these children are subject, urine and stool incontinence is the most frustrating. They have to achieve some forn of evacuation control to be accepted into our society.

During the first two years of life, the incontinence can be managed by the usual methods used for that age group. Beyond that point, however, diapers become socially unacceptable as do accidents of elimination. The full impact of the situation may not be realized until the child reaches school age and finds himself deprived of school attendance solely because of incontinence.

A hygiene program has been developed which is practical and flexible enough to fit any social environment. The program begins in infancy and stresses the early establishment of a socially acceptable method of urine and feces control; special emphasis is put on skin care, fluid and dietary regulation, and awareness of the importance of urinary tract infection. The full understanding and cooperation of the child and his family are necessary and everything possible is done to inform and motivate them.

Bowel Program: Before any type of urine collecting device can be used for either boys or girls, bowel control must be established. [3]

An adequate program consists of (1) keeping the fecal mass of normal consistency or slightly firmer to insure easy evacuation and (2) a timed, consistent evacuation. The bowel program should be established as early as possible, since the bacterial action of the feces and urine complicates skin care. In addition, impacted feces in the rectum presses forward on the urethra to increase retention of urine by preventing proper bladder drainage.

The bowel program is initiated when the child is approximately one year old. One quarter or half of a bisacodyl (Dulcolax) suppository is inserted as high as possible into the rectum every morning before breakfast. The amount of suppository used depends upon the age and size of the child; too much causes abdominal pain, and too little, no action.

The bisacodyl's stimulating action combined with the gastro-colic reflex following a meal produces evacuation in 20-40 minutes. Following breakfast, the child is placed on a pottie chair or toilet with his feet on the floor or a box to flex the hips to the "squat" position. Abdominal pressure is important, so he should lean forward to compress the abdomen against the thighs and/or massage the abdominal muscles. Depending on his age, he can be taught to do this himself. He is encouraged to strain at the same time. For the young child who does not understand the concept of straining, blowing up a toy balloon will serve the same purpose. It is the effort exerted, not just the sitting, that insures success.

The establishment of a daily, consistent routine is essential in maintaining a program. A twice daily regimen may have to be used at first to decrease accidents. Eventually, it is hoped that a daily morning routine can be established. An evening program may interfere with the child's activities, particularly as he enters school. After the program has been established, the use of Dulcolax suppositories is usually no longer necessary.

Occasionally a child may be seen who constantly has diarrhea. This is usually a false diarrhea caused by obstinate constipation or an impaction with liquid feces seeping around the hardened mass of stool in the intestines. Abdominal examination by the physician or nurse will determine if the child is emptying his bowels. If fecal impaction is determined to be the cause, a course of action to clear the bowel will have to be initiated -- stool softeners, enemas, and etc. After this, a consistent bowel program should be started.

By trial and error the family and child can learn to avoid foods which will cause diarrhea or constipation. A normal diet with sufficient liquid intake (approximately 1600 cc) will usually keep the stool firmer than normal which proves advantageous as soiling is less likely to occur.

Undoubtedly, the discussion of a bowel program impresses parents as a difficult long-term program. They need to be constantly encouraged that as with any learning process, time and patience are necessary, as success can't be achieved overnight. There will be accidents as is true with any child. Hopefully as the child grows, he will learn to care for these problems himself. The rewards of a happy and socially acceptable child far outweigh the time and effort spent.

Urine Control: A pediatric collection device for boys and special adaptive panties for girls have been designed to help in the management of urine incontinence once the child is beyond the diaper stage. [2,3]

Male: Urine control in the male is relatively simple as he is naturally endowed with a convenient spigot to which to attach an appliance. Boys, especially if they are ambulatory may be fitted with the Hill Pediatric Male Urinal between 2 and 3 years of age (Figure 1). Bowel control must be established and the diapers discarded as the appliance is unsatisfactory if diapers have to be worn since they do not allow for free drainage.

Results of studies in use of the urinal indicated that 80% obtained good results. [2] Modifications have to be made for those children who have decided lordosis or small or abnormal genitalia. Excessively obese children and non-walkers with an upward pelvic tilt are unable to wear the collector successfully since urine cannot drain upward. However, three wheelchair patients have been able to use the collector with success. Verbal and written instructions in special skin and apparatus care are given to the parents and child. A copy of these instructions is also sent to the nurse in the community so that she can assist the family if any problems should arise.

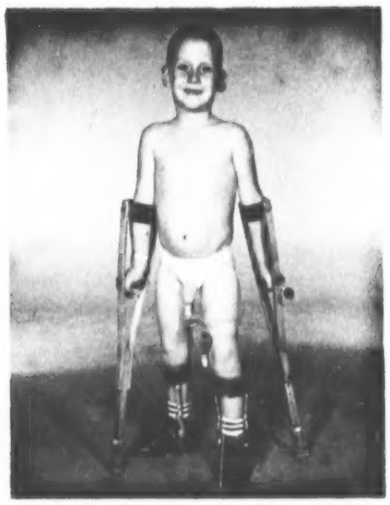

Figure 1.

Hill Pediatric Urinal.

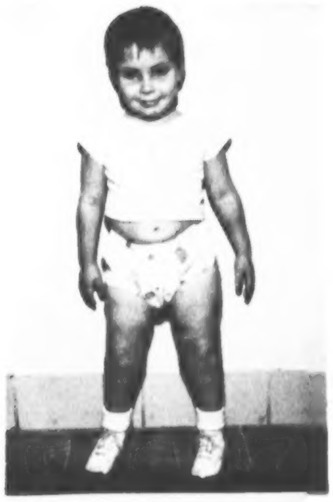

Figure 2.

Adaptive Pantie.

Female: Management of urinary incontinence is somewhat more complicated in girls. At the usual age for bladder training (2-3 years) diapers are changed to "panties". [3] The adaptive pantie is manageable by the child and removes her from the diaper concept. (Figure 2).

For the girls who are able to tolerate the Foley Catheter, the care entails daily irrigations with an antibacterial agent such as Renariden, careful perineal hygiene, periodic catheter replacement, and careful monitoring for infections. However, despite trials of different sized catheters and excellent cooperation of parents and the child. plugging, leakage, and low grade urine infections frequently occur.

Ultimately, most female and male patients, either because of biological deterioration or for social acceptance have an ileal-bladder diversion. The idea is planted as the procedure is discussed with the parents and the child early in their care. More and more of the younger group will ask to have the diversion for social reasons. Only a rare adolescent continues with a catheter, pantie or penile collector.

Skin and appliance care is equally important for the child who has an ileal loop with a collecting bag. This care is demonstrated to the child and his parents during the hospitalization. Both are given opportunities to practice applying the bag as the permanent appliance is fitted before the child goes home.

Detailed written instructions are sent home with the parents at the time of the child's discharge regarding care, application, and where to purchase supplies--and whom to call if there are problems.

By trial and error and experimenting with various appliances, it has been found that the Marlen all-flexible unit* is the most satisfactory. Proper fitting is the key to success. Leakage is usually due to too large a disc around the stoma.

Dilating the stoma with each appliance change is very important. The child and the parents are instructed how to do this during the patient's hospitalization. Here again, the public health nurse insures continuity of care.

Skin Care: Regardless of the type of urine and bowel control in either the male or female myelodysplastic child, perineal skin care is of utmost importance. The combination of perineal anesthesia, dribbling urine and feces, poor circulation and infrequent change in posture, predisposes the sitting patient to rashes and decubiti. The patient and parents need to be taught routine hygiene care from infancy on to provide optimum cleanliness. The habit of frequent posture change should be instilled in the sitting patient. For the ambulator patient, the correct way to sit down needs to be enforced, avoiding a plop down -- the result is a sacral ulcer.

Anti-amonia splitting and neutralizing agents should be used in the care of the diapers or panties for protection and decreasing the amonia odor. Vinegar is still the best and the cheapest.

The management of the multiple defects of a child born with a meningomyelocele requires the collaboration and cooperation of the many specialists in a well coordinated team.

The development of the child as an independent individual is influenced not only by his social environment, but also by his intellectual capacity, the educational opportunities provided for him, his motivation, and the variances of his disease process. The timing of introduction of new skills is as important as the task of cognitive learning. A program initiated for the relief of the problems of these children should be practical and flexible enough to fit into their social environment and enable them to progress to as nearly an independent life as possible. Total involvement and cooperation of all family members are essential in insuring the success of management and the optimal development of the child.

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* Marlen Manufacturing and Development Company., 5150 Richmond Road, Bedford, Ohio 44146

Continued education and reinforcement of the learned processes for both the child and his family are essential. Intensive home visits by the public health nurse are crucial when the care program is being learned. As the families become more independent, less frequent visits are necessary.

In conclusion it may be pointed out that principles used in management of the myelodysplastic child are applicable in those with other disabilities.

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A Bowel Program for the Myelodysplastic Child

Margaret L. Hill, M.N., P.H.N.

Your pediatrician or family physician and/or others undoubtedly has referred to a bowel management program for your child. This does not mean that his bowel can be trained to function normally. It does mean that some form of stool continence can be established by a combination of fluid and dietary regulation, suppositories, and planning a consistent daily evacuation schedule.

At the Birth Defects Center, Univeristy of Washington, our program is initiated at approximately one year of age since an early, consistent training regimen is essential in establishing a daily stool evacuation. Early establishment of the program also makes skin care in the buttocks area less complicated as the combination of chronic dribbling of urine and stool, loss of sensation, and infrequent change in posture predispose the child's skin to rashes and ulcerations.

An adequate bowel program consists of (1) keeping the stool slightly firmer than normal as soiling is less likely to occur than if it is soft or liquid, and (2) a timed, consistent evacuation. Our bowel routine is described as follows:

1. Before breakfast, insert ½ or ¼ Dulcolax suppository as high as possible into the rectum. (Cut the suppository lengthwise with a razor blade. The amount used is dependent upon the age and size of the child--too much causes abdominal pain, and too little, no action.) For older children, you may have to use a whole suppository.

From: Division of Congenital Defects, Dept. of Pediatrics, University of Washington

2. Following breakfast, place your child on a "potty chair" or toilet with his feet on the floor so that the hips are flexed to the squat position. Because of the stimulating action of the Dulcolax suppository on the membrane of the colon and the gastric-colic reflex following a meal, evacuation usually takes place in 20-40 minutes.

3. Have your child lean forward to increase the abdominal pressure by compression against the thigh and massage the abdominal muscles or flex the knees upon the abdomen. Encourage your child to strain at the same time. (For the young child who does not understand what straining or "grunting" means, have him blow on a toy balloon. Children also learn by imitating others.)

The establishment of a daily, consistent routine is essential in maintaining a program. A twice-daily regimen may have to be used at first to decrease accidents. Eventually, it is hoped that a daily morning routine can be established. (We have found that an average program interferes with the child's activities, particularly as he enters school.) After the program has been established, the use of Dulcolax suppositories is usually no longer necessary.

If diarrhea should occur, it is wise to consult your physician, since it may be caused by obstinate constipation or an impaction. (Liquid feces seep around an impacted stool resulting in a false diarrhea.) Your doctor can tell by an abdominal examination if your child is emptying his bowels. He may then recommend treatment.

By trial and error the family can learn to avoid foods which will cause diarrhea or constipation. A normal diet with sufficient liquid intake will usually keep the stool slightly firmer than normal which proves advantageous as soiling is less likely to occur.

Undoubtedly, the discussion of the bowel program has impressed you as a difficult long-term program. As with any other learning process, time and patience are necessary to establish control. As your child grows, he will learn to care for these problems himself. The rewards of a happy and socially acceptable child far outweigh the time and effort spent.

HILL PEDIATRIC URINAL HOME CONSTRUCTION GUIDE

BY

Margaret L. Hill, M. N., P. H. N.

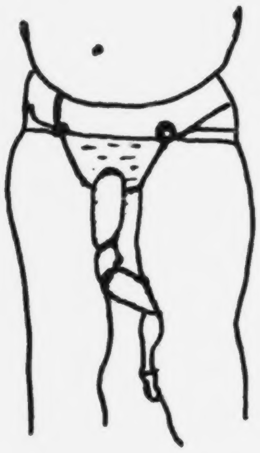

Illustration 1*

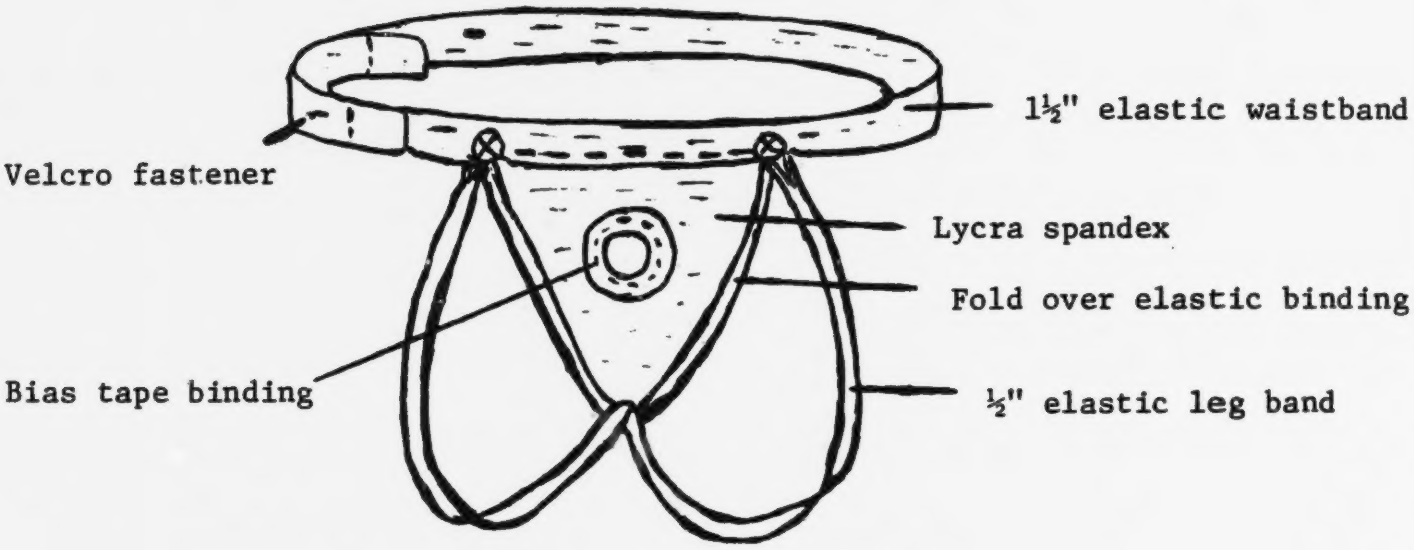
1½" elastic waistband
Velcro fastener
Lycra spandex
Fold over elastic binding
Bias tape binding
½" elastic leg band

Illustration 2

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* The Hill Pediatric Urinal is commercially available from the Davol Rubber Company, Providence, Rhode Island. Parts may be purchased separately. Pediatric latex condom is available in two sizes.

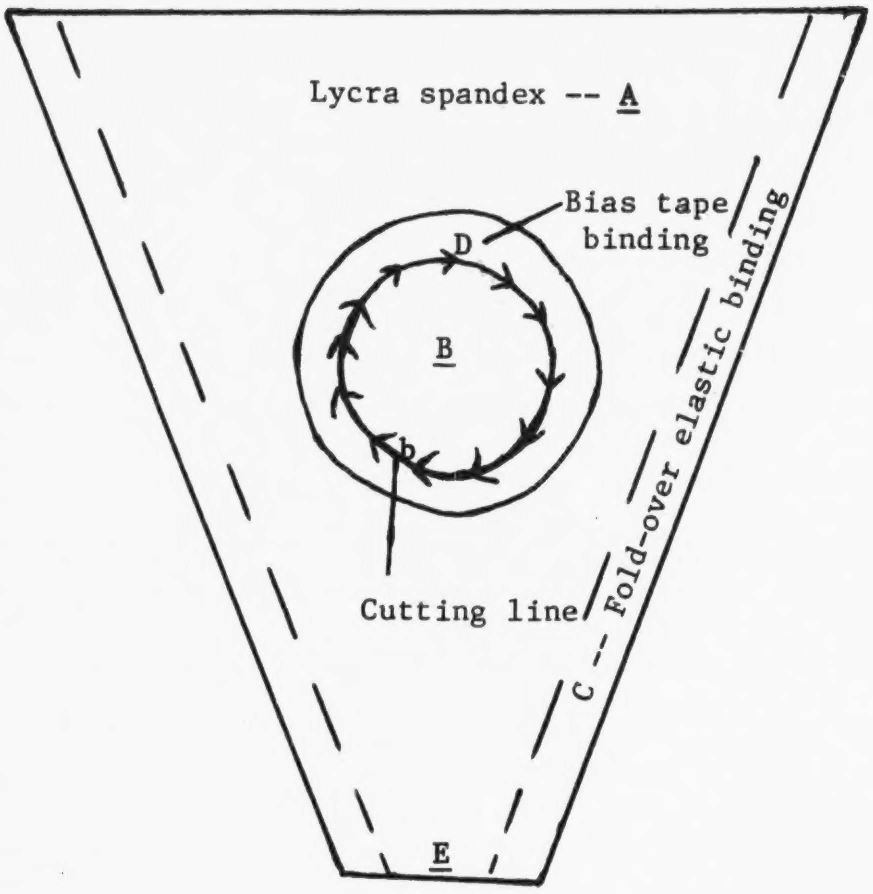
From: Division of Congenital Defects, Dept. of Pediatrics, University of Washington

Supporter Home Construction Guide

Materials:
- Lycra spandex
- 1 1/2" Pajama webbing
- 1/2" Elastic
- 5/8" Fold-over elastic
- Oval elastic
- 2 Buttons
- 2 inches of Velcro
- Bias tape seam binding

All materials are available at department store notion counters.

Pattern Guide:



1. Using pattern guide, cut out triangle of Lycra spandex A.

2. Cut out circle in Lycra using cutting line b as guide. Sew bias tape seam binding around the cut to reinforce the edge.

3. Bind edge C with fold-over elastic binding. If sewing machine does not have a zig-zag attachment, increase length of stitch and stretch Lycra and binding when sewing to prevent thread from breaking when supporter is in use.

4. To obtain length of waist elastic, take measurement over hip bone, (refer to Illustration 1), minus 1" for elastic stretch. Add 2 inches to elastic for Velcro fastener overlap. Sew elastic waist band to A, (refer to Illustrations 1 & 2). Sew Velcro fastener to waist band as illustrated in Ill. 2. Placement of the Velcro is individual to prevent skin irritation or chaffing.

5. To obtain thigh strap measurement--measure from crotch back under buttocks muscle; fold around hip to outer edge of pubic area (refer to Illustration 1), minus 1" for stretch.

6. Double elastic length minus 2" for stretch and sew to E as indicated on pattern. To end of leg elastic strap, attach oval elastic. Sew on button, (Illustration 2).

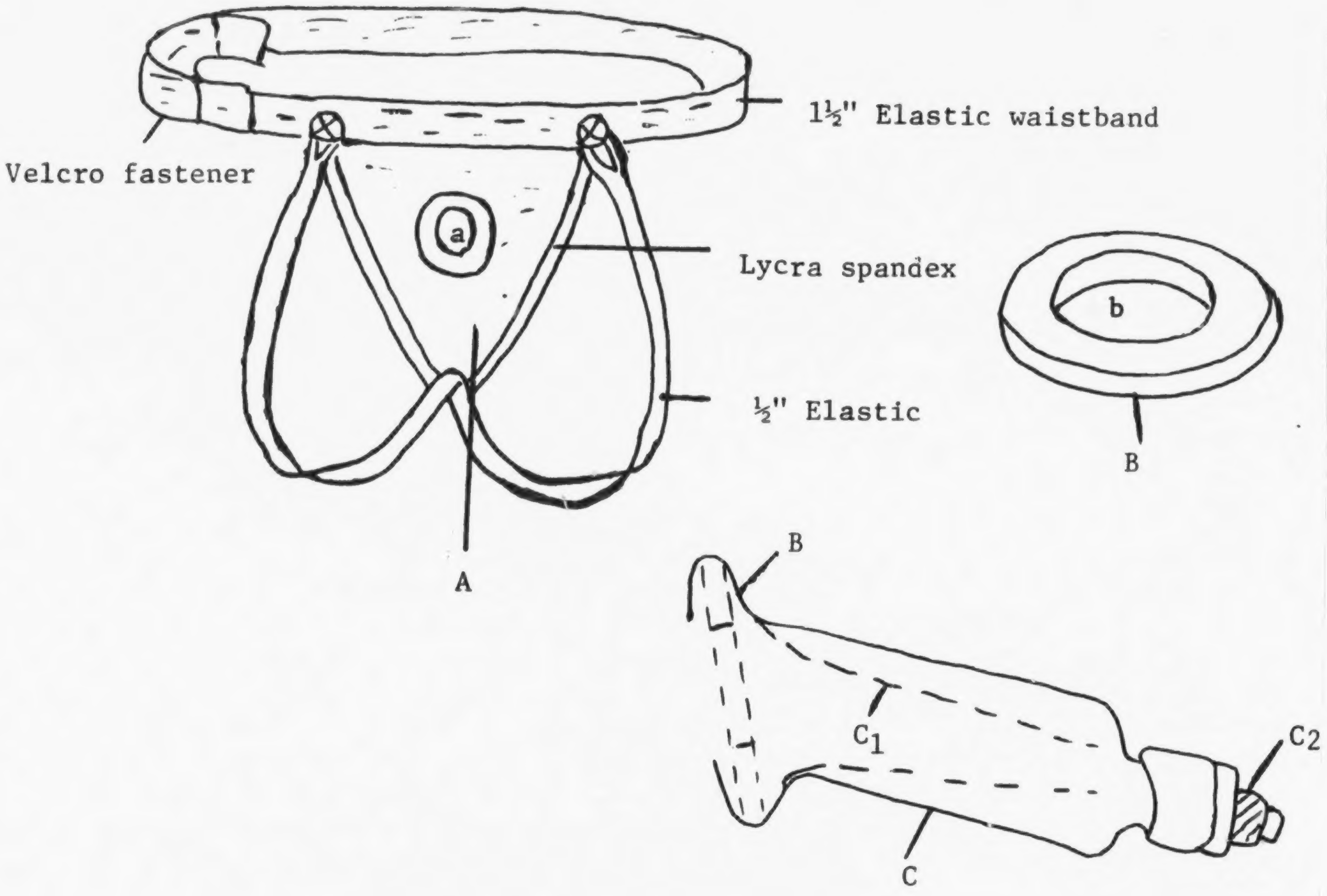


Illustration 3

Directions for use:

1. The Lycra spandex supporter must fit snugly in the pubic area. Individual modifications are often necessary to insure a snug fit. Increase or decrease size of Lycra spandex triangle A, adjust waist band and leg straps so that urinal moves with the supporter as one piece.

2. The latex sheath C is held on to the penis by the insert latex disc B which is incorporated into the supporter through opening a. The penis projects through the latex disc B with the opening cut to penile dimater. The tapering distal end of the inner sheath (C_1) is cut off to the diamter of the penis. This end which is thin and not tight, lies snugly in the coronal groove of the penis. The collection bag (5, 10 and 15 oz. by Davol) screws onto C_2 and is strapped to the inner part of the leg by a tubing connector to the bag, which makes emptying easier for the boys with long-leg pants.

3. The 5, 10 and 15 oz. collection bags are available from the Davol Company. (C.R. Bard, Inc., Murray Hill, New Jersey, makes a 10 oz. male leg bag that is interchangeable with Davol adapter.)

4. Special skin and apparatus care are necessary to prevent penile ulcerations.

   1) The pubic area and genitals should be exposed to the air at regular intervals.

   2) Wash the penis and perineal area and dry the skin thoroughly before applying and after removing the device. Soap containing phisohex is preferred (Dial, Zest, etc.).

3) Wash the appliance and urine collection bag with mild soap and water, dry, and powder with cornstarch. If urine odor persists, soak in vinegar water (4 tablespoons to 1 quart of water) 15 to 30 minutes before washing.

4) Do not use ointments or powder on the penis or perineum, (unless specifically ordered by the physician), as they predispose the area to ulcerations by localizing the bacteria. If penile ulcerations occur, discontinue use of the urinal until the skin is clear.

5. Two collection devices should be available to use alternately.

6. The appliance is not recommended for night use as the child rolls in bed and pulls the urinal off the penis, resulting in leakage.

7. The appliance is unsatisfactory if diapers have to be worn, as they do not allow for free urine drainage. Special modifications are often necessary for children that are non-walkers or have lordosis (sway back). The penile opening in the supporter may be placed posterior to fit the base of the penis. Often some type of skin adhesion can be used to adhere the appliance and decrease leakage.

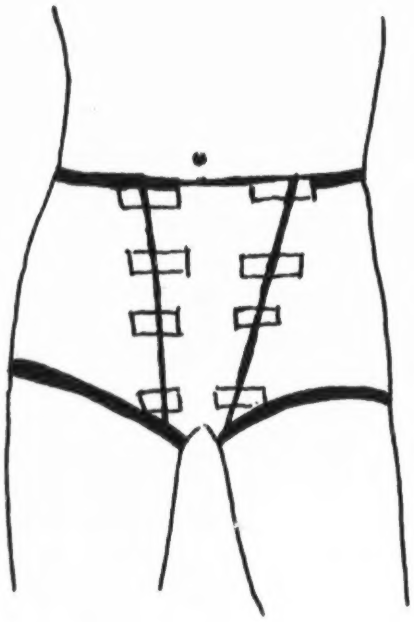
8. For emergency use, the medium-sized Lapides condom may be substituted for the Davol urinal C. Cut out a thin latex disc resembling B with the hold b to fit snugly over the penis. Refer to #2 for directions. Attach the end of the condom to an adapter to which the collection bag may be attached.

9. As each patient presents a different situation and different problems, the recommendations will need to be adapted to his particular needs.

THE ADAPTIVE PANTY HOME CONSTRUCTION GUIDE

by

Margaret L. Hill, M. N., P. H. N.



The adoptive panty allows the incontinent child to be free of bulky diapers. A plastic pocket-like moisture-proof inner liner extending from the waist front through the crotch to waist back holds the incontinent pad in place. These pads are easily replaced without having to change the entire panty. Because of the front opening, children who have braces can put on and take off these panties without having to pull them up over the braces.

Directions for Home Construction:

Materials

Any washable material such as terry cloth, cotton jersey, nylon tricot, or cotton broadcloth.

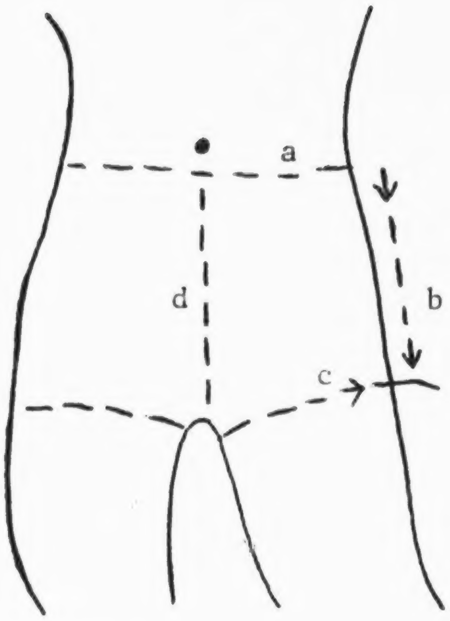
Snaps, buttons or velcro for front fastners.

Twill or bias tape to finish edge.

Plastic (boil proof) for interliner.

1/2 inch elastic.

From: Division of Congenital Defects, Dept. of Pediatrics, University of Washington



1. Take body measurements as indicated in Illustration 1.

   a. Measure waist.

   b. Side length - measure from waist ↓ 7 inches.

   c. Thigh - measure upper leg at thigh (7" point ↓ from waist).

   d. Overall length ↕ measure from waist back through crotch to waist front.

Illustration 1

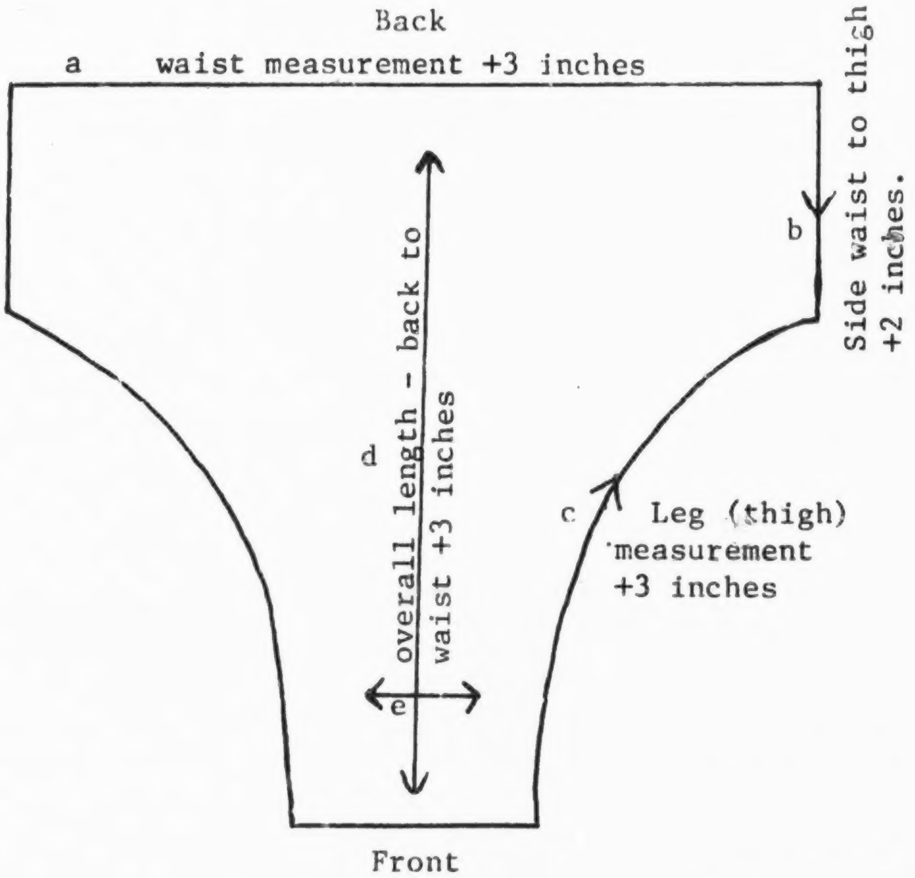

Back
a
waist measurement +3 inches
Side waist to thigh
+2 inches.
b
d
overall length - back to
waist +3 inches
c
Leg (thigh)
measurement
+3 inches
e
Front

Illustration 2

2. Make basic pattern as outlined on left. To allow for seams add to:

Waist measurement	+3 inches
Side measurement	+2 inches
Thigh (leg) measurement	+3 inches
Overall measurement	+3 inches

The overall length d ↕ should not go higher than the waist (preferably slightly below the waist) or the panty will not fit snugly. The width of front ←e→ should be wide enough so that panty fits snugly against inside thigh.

3. Cut out panty from desired material.

Insert lining -

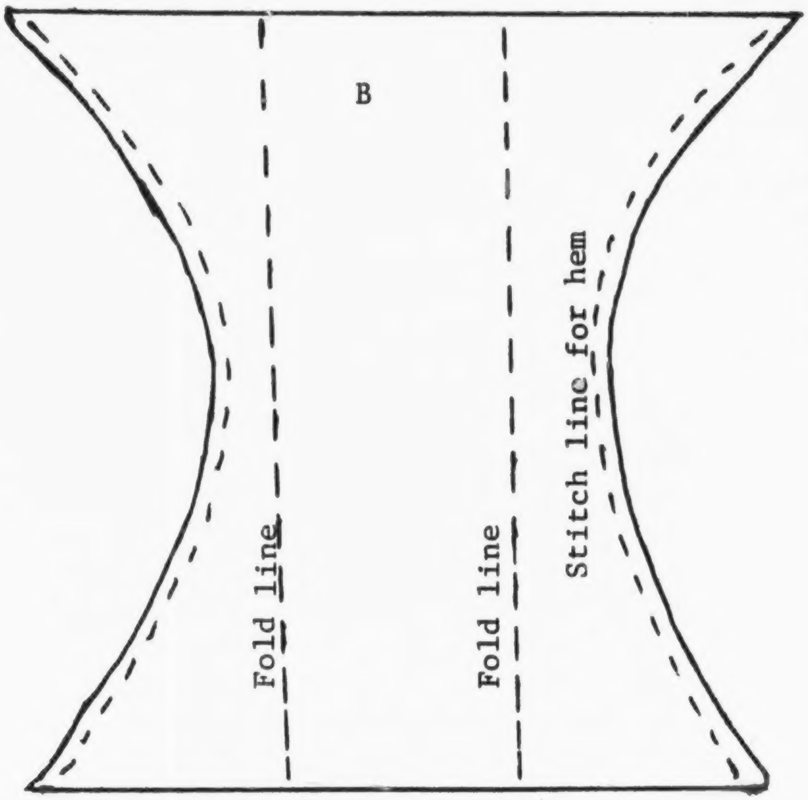

B
Fold line
Fold line
Stitch line for hem

Illustration 3

4. Cut 1 plastic insert B. Turn outer edge (as indicated) and make 1/4 - 1/3 inch hem. (reinforces plastic so won't tear)

(Medium weight boil-proof plastic is recommended and is available at most department stores.)

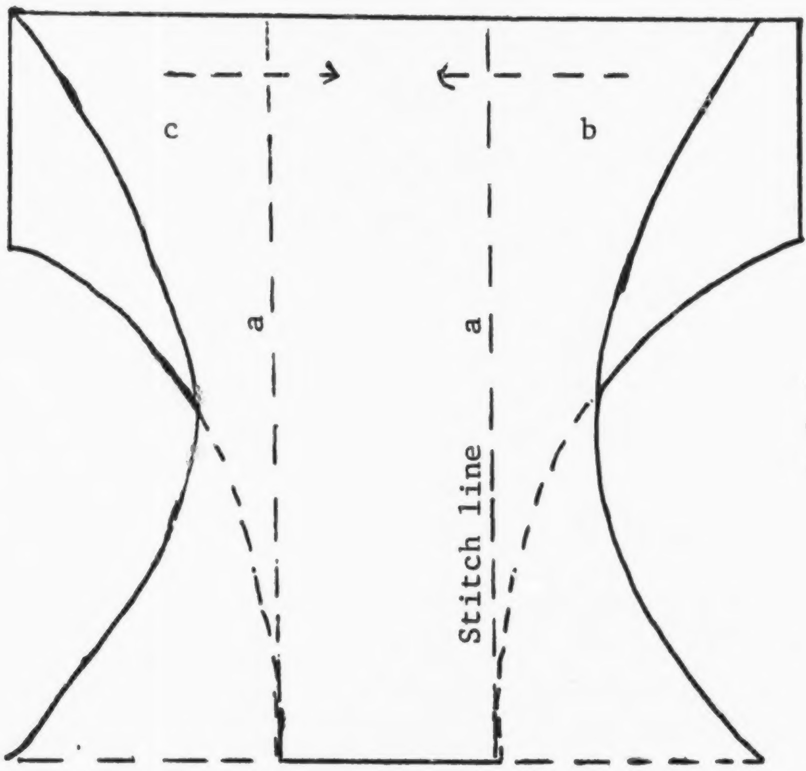
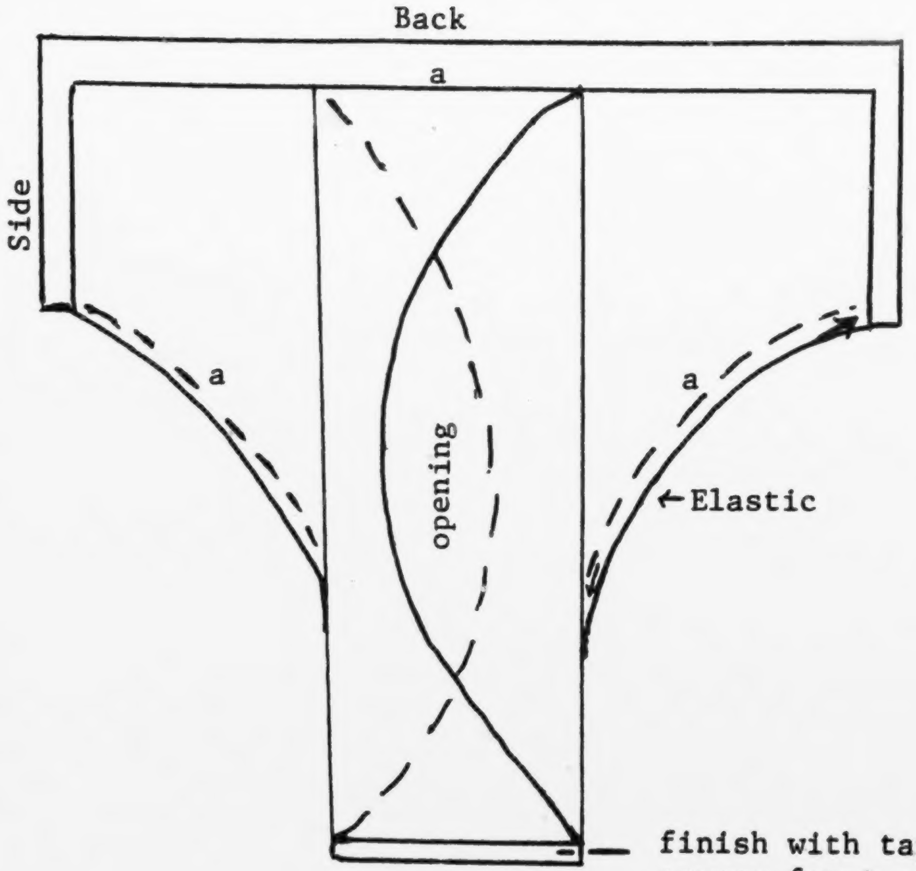

c
b
a
a
Stitch line

Illustration 4

Place plastic insert B on inside of panty. Stitch along stitch line a. Fold flap b in over stitch line. Next fold flap c in as indicated in Illustration 4.


Back
a
Side
Side
a
a
opening
←Elastic
Front

Finish edge a of panty with twill or bias tape (3/4"). (Leave opening to insert leg and back elastic.) Insert 1/2" elastic and gather up slack so that panty fits snugly. Allow slack for fastners.

finish with tape but do not insert elastic across front piece.

-Illustration 5

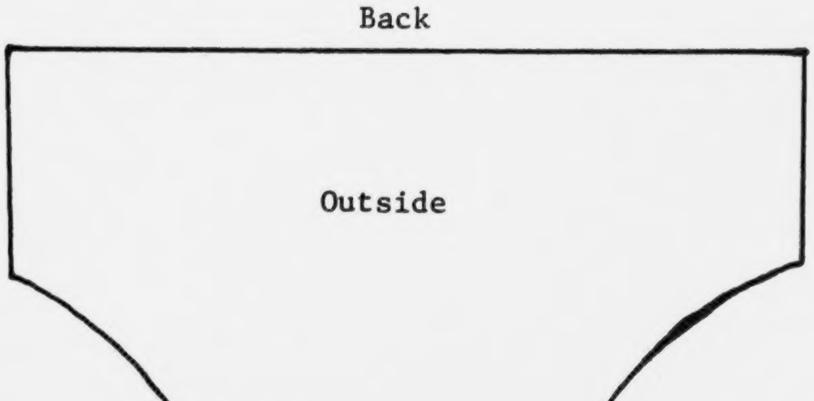

Back
Outside
Front

Illustration 6

On outside front piece attach strips of hooked teeth velcro as indicated on Illustration 6. Be careful not to sew through inside plastic insert pocket as you will not be able to insert diaper pad.

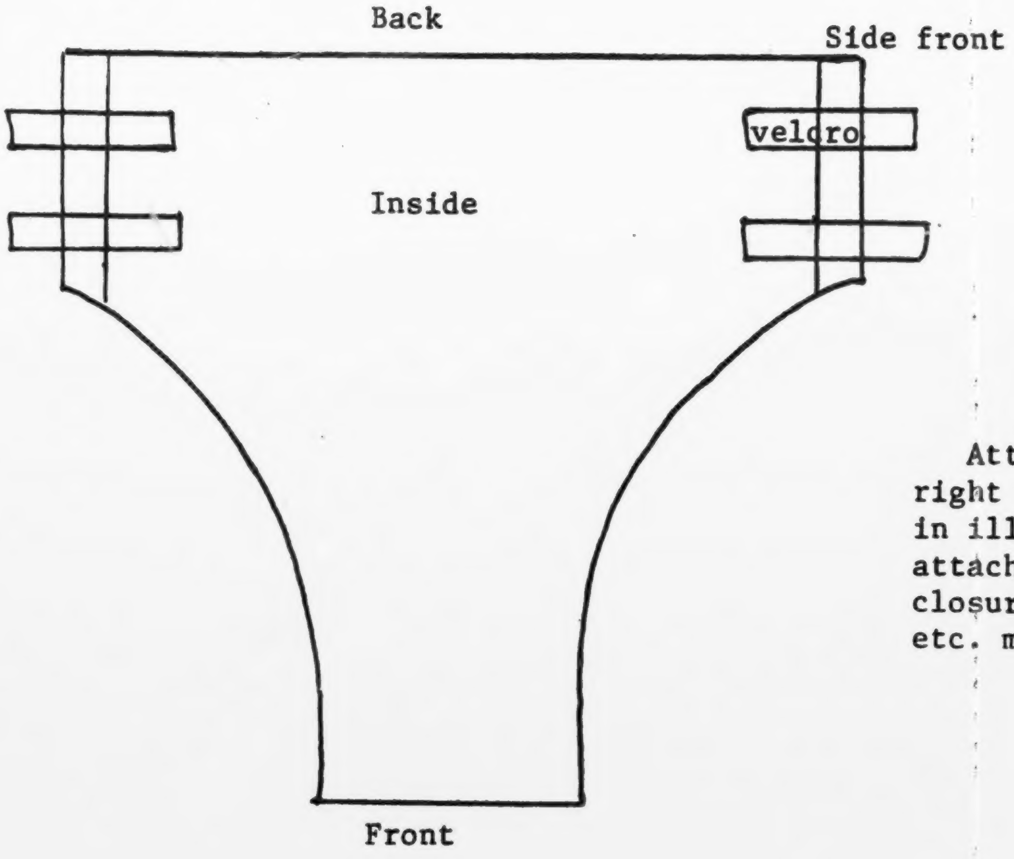
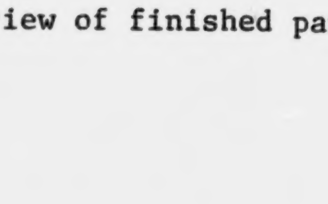


Illustration 7

Attach strips of soft velcro to inside right and left side of back as indicated in illustration, when complete panty attaches in front. Various types of closures such as buttons, snaps and etc. may be used.



Front view of finished panty.

As each child presents a different situation and problem, the pattern and recommendations for making will need to be adopted to his situation and problem.

THE OLDER CHILD

EDUCATIONAL IMPLICATIONS FOR THE SPINA BIFIDA CHILD

Betty Green, Ph.D.

The child with spina bifida has not only the needs and problems of childhood but also special kinds of needs and problems, both physical and psychological.

Experience has revealed that the degree of the handicap cannot be related proportionately to the exact degree of physical limitation or disability. The term "handicapped" must be refined to speak of the "functional limitation" of the spina bifida child. The actual disability is dependent upon the severity of the defect. Teachers, nurses and therapists should be aware of the actual disability so that they will know the basic physical capabilities of the child.

In working with spina bifida children, it is apparent that one of the most difficult tasks they face is personality adjustment. It is heartwarming to see a child with a friendly, cheerful disposition and a zest *for a good adjustment to their* way of life and a means of adapting to what could be great frustrations. Cruickshank, on the other hand, found in a study that the physically handicapped group as a whole shows a greater tendency toward a personalized introspective view of life with concern over the effect of the disability. Many crippled children have difficulty facing social situations and those which may imply guilt or personal inadequacy. So indeed one important factor in dealing with this child has to do with the child's acceptance and understanding of his own handicap and the acceptance of him and his disability by his family and the people around him. Because society has placed a premium on physical beauty and ability, it is easy for a crippled child to devaluate himself. A family which places great stress upon athletic ability and prowess will have difficulty accepting a boy with a spinal defect, braces, crutches or a wheelchair. However, a family in which intellectual skills play a large role may well be able to accept the child and allow him to compensate for his physical deficiencies by furthering the development of his intellectual abilities. Those children whose disabilities are not accepted are actually often unwilling to go along with remedial measures in their attempts to deny the seriousness of their problems.

Being accepted, loved and wanted all lead to the sense of security of belonging, and this is something that every child needs--and the spina bifida child may have more difficulty in acceptance if he carries the stigma of an odor. Teachers and the school can aid in this respect by fostering the things that a child with spina bifida can do as well as recognizing the things he cannot do. There are social activities, organized play and various interests and hobbies that he can develop in addition to his educational achievements.

There are many problems which the child must work out in his relations with the outside world, such as the difficulties in sources of recreational activity compatible with the limitations imposed by his handicap and in achieving recognition for his successes.

It may be easy to recognize and understand the handicapped child's problems in dealing with the outside world, but it is more difficult to conceptualize and understand his internal psychological problems, and how the two often intermingle. It is important to establish such differentiations since help, and sometimes schooling, for these children are often not used effectively because they deal specifically with the external struggles. Because of the nature of the child's disability and his inability to get out and do things as easily as other children, his dependency upon others is prolonged. This creates a great struggle within the child. Because of this tendency to cling to the protective environment longer, this struggle is caused by the thoughts the child himself may have about the handicapped parts of his body or their limited functions. His ideas and feelings may bother him to the extent that he may try to rid himself of them either by denying the presence of the handicap (whether it be paralyzed legs, or bowel and bladder incontinence) or he may fail to grow up emotionally.

Even though the handicap has existed from birth, the child senses his handicap, sometimes acutely, for the first time in the classroom. Therefore it is important to help him become accustomed to being with other children and to accept his wheelchair or braces as a part of himself. Although the educational program for such children often needs to be changed to meet his needs, it is vital to provide normal and rich educational opportunities, varied social relationships, and recreation.

Educators and others should remember that the other children need some help in understanding and adjusting to him. Many people involved with the teaching of such youngsters have found that mixed classes of physically normal and handicapped children can be of great benefit to both.

Spina bifida children when faced with intolerable frustrations may become aggressive, blame other people, withdraw into fantasy, regress by using childish behavior or become very demanding, or they may compensate for the disability by finding an alternate interest which will satisfy the same motives. Compensation is easier for us, and we all do compensate at one time or another, because we have a wider range of abilities and activities on which to draw, but this child needs to find an area of satisfaction in which he can compensate (could be art, music, debating, chess, etc.). Once they achieve success, they become more objective toward their physical handicap and are willing to accept it. Many children can compensate by working for high academic achievement, but since this is not possible for all of them, some may gain satisfaction from social and personal achievement. All that can really be said about this is that attempts should be made to determine the areas in which these children can really achieve to their own satisfaction.

When at all possible, the spina bifida child should be integrated into the regular classroom. They should have the opportunity to have as much contact as possible with the non-handicapped child. In some cases adaptations may have to be made in the physical plant as wider doorways, ramps, and in some instnaces, elevators. Whenever possible, the handicapped should be encouraged to participate in school activities like assemblies, lunch periods and certain playground activities. A spina bifida child with a degree of mental retardation may possess a particular problem but generally the handicapped child should be allowed to progress academically in the regular class if at all possible.

Teachers should always be consulted before placing an orthopedically handicapped child in their class. The teacher needs to be one that can set aside her overly sympathetic attitudes, and particularly any tendency toward unrealistic thinking. Teachers will find, though, that many orthopedically handicapped children have special, legitimate emotional needs which must be met before progress can be made toward achieving educational goals. A teacher of the handicapped should possess all the qualities essential to being a good teacher of normal children. If the classroom teacher has any physical limitations of her own, this should be considered though, if there is any lifting of these children involved.

If at all feasible a teacher's aide should always be placed in a room that contains handicapped children. A spina bifida child may not only need assistance using the restroom facilities but with putting on his coat, getting out of the chair, or tying his shoelaces. All of these duties that can be taken from the teacher gives her more time in the actual work of teaching. A teacher can be both physicallyand emotionally worn down if she has to plan, direct, and actually teach a whole classroom of children with the added burden of time-consuming non-teaching problems that are encountered in having physically handicapped children in the room. A spina bifida child may require even more time than some children and a teacher may be more willing to have this type of child in her room if she knows she will have assistance.

It is much more beneficial to the child if he can be in the regular classroom, rather than on homebound care. At home he receives no social contacts, no self-help skills, no outside influences and sees his teacher only one hour a day.

The primary educational goal for spina bifida children is identical with that for any other group of children: to enable them to become contributing members of society. One way to accomplish this task is to coordinate school and home activities by establishing good communication between parents, teachers, therapists, psychologists, and doctors. This is important for the parents to learn how to help the child accomplish these goals in purposeful activities outside the classroom. Contacts between the home and school can be accomplished in many ways--scheduled conferences, phone calls, and written communications. The teachers also can make home visits.

It is important that teachers and administrators of special education programs establish positive and strong lines of communication with parents and help them understand and accept their child. Psychologists, medical consultants, and counselors should be used to advantage in the parent counseling program. Regularly scheduled and well-planned parent-teacher conferences can be a most effective medium of communication. Parent-teacher organizations can be most valuable in promoting home and school cooperation. Even though some parents may find it difficult to attend these meetings because of wide distances to travel, the programs should be planned to be so rewarding in terms of parent needs that they will feel they cannot afford to be absent.

Need for parent counseling is exemplified by a study made in Iowa. One hundred-fifty parents of cerebral palsied children were sent questionaires; results might be comparable to responses from parents of spina bifida children.

52%---thought the doctor was responsible for their child's cerebral palsy
69%---expected their child to get a job eventually
71%---expected them to marry
49%---expected their child to be completely cured
100%--of the parents thought physical therapy would be helpful but
    40% thought that they did not have time to carry out this treatment at home
62%---thought their child was sent to them as a punishment
70%---thought that God had given them their child as a cross to bear
91%---were afraid to have other children
48%---of the parents did not know that brain injury caused cerebral palsy
80%---thought that surgery would help their child

Parents need help in arriving at a common sense understanding of the facts about their children, what can be expected from physical therapy, occupational therapy, and education, and what the future is likely to hold. Through lectures and demonstrations, especially those followed by discussions, many parents are able to learn facts and to acquire attitudes which help them better accept their child and plan more realistically.

The attitudes which have been prevalent in the home setting will of course determine the kind of response the child will have in school. The expression of aggressive impulses which were discharged formerly by any means must now be expressed by verbal and intellectual methods. This makes the teaching of the spina bifida child different from teaching the normal child. Much skill, versatility, and patience are required. In a school where the child can be perceived accurately, and where he himself gains an accurate appraisal of his own limitations together with his own capacitites, such a child may emerge psychologically healthy.

RECONSTRUCTION OF THE HIP IN SPINA BIFIDA

W.J.W. Sharrard, M.D., F.R.C.S.

Dislocation of the hip is present at birth or develops during the first three years of life in more than half of all children with meningomyelocele. When a child presents with dislocation of the hip at birth, it has been tempting in the past to try to treat it as one would a congenital dislocation of the hip. In fact, the pathology and the whole position of the hip are quite different in spina bifida from that in congenital dislocation of the hip.

When the hip is dislocated at birth, there is always severe fixed flexion and adduction deformity, something quite different from that in ordinary congenital dislocation of the hip. Examination of the muscles shows that there is always strong action of the hip flexors and adductor muscles and severe or complete paralysis of all the gluteal musculature. This implies that the dislocation is a paralytic one. It has occurred in utero and is not so much an associated deformity with spina bifida but a secondary deformity arising from neurological loss.

When the dislocation occurs later, it is still found that the lesion is a paralytic one. In this instance, there may be a little subluxation at birth which leads to dislocation at about the sixth month. In these patients, there is innervation down to the fifth lumbar segment with preservation of some gluteal function but not sufficient to prevent the hip from gradually becoming more flexed and adducted and eventually dislocating. Another way in which dislocation may develop is when the spinal lesion is treated late or infection occurs in the plaque, so that a child who previously had normal innervation in utero becomes secondarily paralyzed in the sacral segment. This will also result in dislocation after one to two years. Then there is the problem of the child who has innervation down to the first sacral segment. The hip does not tend to dislocate but flexion deformity is likely to develop because of weakness of the gluteus maximus. Finally the child who has complete paralysis of the hip tends to have his hips in abduction and external rotation into which position they tend to roll when he is lying down.

Before discussing treatment, a word needs to be said about the pathology of the lesion as regards the bones and the condition of the hip. In the dislocated hips, even when there has been intra-uterine dislocations, there is a remarkable preservation of a good acetabulum. I believe that this is because the hip dislocates and the acetabulum continues to form even though the head of the femur is not in it. The acetabulum may become deformed in patients in whom the hip is in joint at birth but starts to subluxate during subsequent months. The mechanism of this is, I believe, one in which the axis of abduction and adduction of the hip moves down from the centre of the head of the femur to the region of the lesser trochanter.

It does so because the psoas becomes tight and in effect the hip is swinging on the psoas so that the head of the femur is moving in and out of the acetabulum, in abduction and adduction. A further result of this is that the roof starts to become worn and eventually a typical acetabular dysplasia develops. With regard to valgus of the neck of the femur, it is always associated with ante-version and it is due to weakness of the abductors. Of itself it does not give rise to dislocation and if a child has a completely flail hip he will not dislocate even though the neck of the femur is in very severe valgus.

CORRECTION OF DEFORMITY

In earlier days in these children, we tried to reduce the dislocation at birth but this proved to be impossible. Attempts to reduce the dislocation and to apply a splint invariably result in failure. If one does succeed in obtaining abduction it is usually because the child has sustained a fracture of the upper third of the shaft of the femur. Passive movements by physiotherapists are equally likely to fail and may be disastrous. Operative correction is not feasible in the early days of life because of the other lesions that are present.

I believe that it is unsafe to try to correct deformity of the hip by any conservative means. Pressure sores, circulatory disturbance even leading to the need for amputation, and fractures are very likely to occur. The only safe way to correct the dislocation of the hip is by correction of the deformity which means elongation or division of tight muscles. The tight muscles are the adductors and the psoas.

The adductor tightness can be released by adductor tenotomy. This should never be a subcutaneous tenotomy which is either inadequate or dangerous. Open adductor tenotomy should be performed and it is usually necessary to divide all the adductors completely. The aim should be to gain 90 degrees of abduction with preservation of the obturator nerves. If this is done, in many patients the hip dislocation will reduce. It is important not to retain the position of full abduction in flexion, however. If this is done the legs will remain in the frog position indefinitely. The next step should be to release the psoas but it is best to do this at the time when the psoas is transplanted.

The reason for transplantation of the psoas is to correct muscle imbalance. If it is not done, the hip will invariably redislocate because of the pull of the iliopsoas in the presence of the paralyzed glutei. Posterior iliopsoas transplantation will correct this situation. It is best done when the child is between seven and twenty-four months of age. Ideally it should follow within a month of adductor tenotomy and reduction of the dislocation. The psoas tendon is exposed by going between the femoral vessels and nerves and it is essential that the lesser trochanter should be visualized so that the tendon may be divided together with a small piece of the lesser trochanter. It is mobilized up into the pelvis and the whole of the iliacus from its attachment to the inner side of the pelvis. If this is not done, it will be impossible to transfer the tendon to the greater trochanter. A hole is made in the ilium just lateral to the sacro-iliac joint and the whole of the iliacus and the combined iliopsoas tendon passed through a bony tunnel in the greater trochanter. An extremely useful instrument to perform this manoeuvre is a tendon passing cannula which can be made from any ordinary metal sucker such as is used by anesthetists. When the psoas has been detached the hip will then go fully into joint and will lie in abduction, extension and medial rotation. It is important that the transplant should be made as tight as reasonably possible. Fixation is maintained for three or four weeks if the hip has been subluxated and four to six weeks if it has been dislocated. If there is looseness of the capsule of the hip, it is a good thing to reef the capsule but I have seldom ever found it necessary to remove a limbus or to open the acetabulum in a child below the age of two. Additional procedures such as varus osteotomy or acetabular reconstruction are not usually necessary in children below the age of two.

In an older child, that is one presenting with a dislocation of the hip aged four to six years or more, it is often still possible to reduce the dislocation by the same method as that described, but it will now be more often necessary to perform some kind of acetabular reconstruction. It is quite appropriate to perform a Salter innominate osteotomy at the same time as the psoas transplantation because the exposure is all available.

Where flexion deformity develops with paralysis of the gluteus maximus, a posterior iliopsoas transplantation is still sometimes useful though the transplantation should be made to the middle of the shaft of the femur posteriorly rather than at the back of the greater trochanter in order to make the psoas transplant be an extensor rather than an extensor and abductor.

In some children, various kinds of deformity may not necessarily require psoas transplantation but there may be need for various kinds of rotation, varus or even valgus osteotomy. The consideration here should be the placing of the limbs in a position in relation to the trunk so as to allow calipers to be fitted; this is particularly true ot those with flail lower limbs.

In a long term follow-up over 11 years, we have found that if a satisfactory reduction was obtained, the psoas transplant succeeded in maintaining the reduction. In only one case has there been a redislocation nine years later and even this was able to be reduced once more and the tendon transplant tightened. Even where the hip was not able to be reduced originally, posterior iliopsoas transplantation on the dislocated hip has seemed to be worthwhile. Seventy-five per cent of children have become able to walk without braces at hip level. The power of the transplant has been found to be power 3 or more in 17 per cent and power 2 to 3 in another 30 per cent. In the remainder, no definite active abduction can be demonstrated but the child is able to stand upright and the transplant is presumably acting as some kind of muscle tenodesis. Up to the present time we have performed about 750 posterior iliopsoas transplants and we have found no reason to alter this policy or even to alter the technique which seemed to come out right from the very beginning. We have, of course, tried several other ways of doing the operation to see if it could be made easier, particularly such modifications as altering the approach to the lesser trochanter. Personally, I still find the approach between the femoral nerves and vessels the best, even if at times it is a little bit tedious.

An important consideration after the child comes out of plaster is the possibility of spontaneous fracture of the lower end of the femur. If it occurs, there will be no pain but it will be shown by swelling of the limb. Union occurs very promptly. It is worth noting that these children do suffer from defective Vitamin C absorption and this may be a factor in the liability to fractures. All spina bifida children should, therefore, be given large quantities of additional Vitamin C and great care should be taken during the first two or three weeks after the plaster has been removed to avoid unnecessary trauma to the limbs. After this time, physiotherapy is started for those who are old enough to learn how to walk. We sometimes use full braces to support the hips at first but soon graduate to long leg braces and later to below knee supports which is all that these children should need.

THE INDISPENSABLE NURSE

Margaret L. Hill, R.N., P.H.N., M.N.

Who is she?

> She opened her hands to the needy and stretched out her hands to the poor...Strength and honesty are her clothing...She rendered good, not evil, all the days of her life...The law of clemency is on her tongue... Many daughters have gathered together riches, but the valiant woman has surpassed them all.
>
> The Book of Proverbs

She is a member of a health team working with other disciplines to provide optimum care for the child and his family. She sees the child as an individual, a member of a family and a community. She combines her philosophy with her scientific knowledge and from this orientation exercises sound judgment in directing and giving nursing care. This care relates to the patient's whole life: his family, his home, his place in the community, and his self-concepts as a person. It includes those activities which she performs directly with the patient and his family, and those performed away from the patient and his family, but in their behalf.

What then is the unique function of the nurse? It is the identification or diagnosis of the nursing problem and the implementation of nursing action for the solution of the problem. However, once she has identified the problem or need, she then develops her plan of care in cooperation with the health team members. How the nursing care plan or action is implemented is dependent upon the environment in which the nurse functions. Continued evaluation of the care plan and allowing for change are also essential components of the nursing process.

The Nurse in the Clinic and the Community

Increasing emphasis on comprehensive medical care coupled with rising hospital costs have stimulated hospitals and clinics to recognize their responsibility does not end within four walls. [1,2] The home environment (even if far from ideal), family and friends provide the patient with security and love, which are essential components in the restoration of maximum health. [3]

However, total care in the home can be appropriate only if the provision for continued comprehensive medical care has been adequately assured by an efficient system of coordination. To insure clear communications and participation essential for comprehensive services, the nurse member of the team should be in a central coordinating position. With the help of one physician in charge of the continuum of outpatient and inpatient care, she assists the family with recommendations of the members of the medical center health team, and provides direct patient care. She also is responsible for coordinating activities associated with the provisions of continuing nursing care between hospital, clinic and patient's home.

All the nursing functions described herein are not appropriate to each clinic or hospital due to differing interests, needs, finances, personnel and communities served. Neither is it possible to implement all the identified functions at one time. As personnel become familiar with concepts, they assume new and different functions peculiar to their own community needs. One essential consideration should always be continuing flexibility to allow for change.

The author of this paper assumed a nursing coordinator-consultant position to the Division of Congenital Defects, University Hospital, Seattle, over a five year period. The primary objective was to define how patient care could be better implemented. To do so, she participated in primary patient care and developed a pattern of nursing participation best illustrated in the following case example.

K.F., age 5, born with meningomyelocele, was referred to our Birth Defects Clinic by the family physician. The family had just moved from out of state. The presenting problems were: (1) partial paraplegia and bilateral dislocated hips which required bilateral long leg braces with a pelvic band; (2) stool and urine incontinence; (3) frequent urinary tract infections; and (4) pending school deprivation because of urine and fecal odor.

1. Coordination of Patient Care in Clinic:

    1. A community public health nurse's (PHN) assessment of the child's function in home and community was obtained prior to her clinic visit.
    2. The public health nurse's home evaluation was reviewed for completeness from a nursing viewpoint. (If pertinent data is missing, the PHN should be telephoned for clarification).
    3. The referral and PHN assessment of needs and problems were discussed with the pediatrician to define a course of action.
    4. The pediatrician ordered a diagnostic evaluation to include psychologic, social service, urologic, orthopedic, neurosurgical and pediatric evaluation to be summarized in a case conference.
    5. The parents, public health nurse, school representative, and family physician were invited to attend the multidisciplinary clinic conference. Participation in a clinic conference by those involved with continuity of care in the community is encouraged. However, because of time and distance this is not always possible as it was in this case.
    6. The nursing coordinator interviewed the family during the complex initial evaluation and assisted them by explaining various procedures and appointments. At this time she obtained supplementary information about the patient's self - care, ability and needs.

7. The coordinator participated in planning the home care program during conference in two ways:
   (a) Presentation and discussion of thelocal PHN's report, community needs, facilities to aid in arriving at a care plan. (If community PHN is present, she presents her own evaluation.
   (b) Discussion of nursing methods to meet overall goals of patient function. In this case the importance of bowel and bladder management and appropriate modifications of our routine were discussed with the group. [7]
8. Following the case conference which decided against surgical diversion of the urine, the coordinator instructed the child and parents in nursing care techniques which included bowel and bladder management, [7] special skin care [7] and the importance of fluid intake, preparation of the child for pending hospitalization and orthopedic surgery, the meaning of the public health nursing referral and community services available for a handicapped child (i.e., Elks Physical Therapists, Crippled Children's Services, Special Education Classes through the school district, etc.)
9. The written referral for public health nursing follow-up was initiated and sent by the coordinator to the Public Health Department describing the home nursing care and the overall medical plan. A copy of the referral was also sent to the private physician, and one was incorporated into the patient's clinical record.

II. Coordination of Patient Care on the Hospital Ward:

To increase K's mobility, orthopedic surgery to stabilize her hips was scheduled as a result of the case conference. The nurse coordinator assisted in the following ways:

1. On admission to the Pediatric Ward, she consulted with the head nurse to insure that the home care plans were continued (suppositories for bowel management, urinary bladder credé, adequate fluid intake). Special hygiene established at home should be continued on the ward, since inactivity following orthopedic surgery predisposes a myelodysplastic patient to constipation and urinary tract infections.
2. The coordinator consulted daily with the head nurse and/or staff nurse responsible for K's care regarding her progress. She contributed to the nursing care by her knowledge about the child and her family. (i.e., parent-nurse and parent-child relationships, anxieties, activities of daily living). At weekly ward rounds with the multidisciplinary staff, the nursing coordinator presented problems of the nursing staff.
3. The nurse coordinator attended the pre-discharge conference and contributed solutions to the special nursing care problems (i.e, special cast and skin care of a child who is incontinent and has loss of sensation in the lower extremities, importance of

weightbearing post operation, plans for home care in a cast, improvised equipment such as a tilt board for standing, and play therapy suggestions for a child who is immobile).

4. The written referral to the Public Health Agency for nursing follow-up was initiated by the ward nurse, after a review and discussion with the coordinator.
5. Upon discharge, the parents were given an appointment for the patient to return to the Congenital Defects Clinic to have the cast removed,to be measured for new braces, and to initiate a physical therapy program. Prior to this clinic appointment, a progress report was received from the public health nurse. The progress of the patient at home was uneventful as reported by the public health nurse. After it was reviewed by members of the health team, the referral was incorporated into the patient's permanent record.

DISCUSSION

The coordinating nurse can fill a unique professional role in coordinating hospital, clinic and community services, developing comprehensive nursing care plans, and providing continuity of care. These functions can be most easily delineated as : patient care, teaching and research.

Patient care: Her primary area of function should be patient care. Direct involvement enables the coordinator to practice current nursing techniques. It also allows her freedom to improvise and develop improved nursing care techniques and to improve her own clinical competence.

Indirect care may be described as those activities carried out collaboratively with other nurses and/or professionals. The use of an efficient interagency referral system to extend comprehensive care is an example of indirect care.

Not all patients will need a nursing referral for home care. Dr. Louise Smith [6,2] suggests certain criteria in assessing the patient's need for a nursing referral:

(1) A procedure that requires professional assistance in the home
(2) A need for reinforcement and clarification of instructions started in the hospital
(3) A patient and/or family unable to accept or are disturbed by some aspect of his condition
(4) A physical or social environment at home which may interfere with the patient's self care
(5) A need for anticipatory guidance for mothers with young children and particularly for parents of handicapped children who may have deviant patterns of development.

A public health nurse home evaluation assists in assessing the need for a nursing referral. If a referral is appropriate, the patient and his family should be advised as to its meaning, and how the nurse can be expected to help. In addition, the need for nursing referral should be discussed with the family physician by a physician member of the medical center team.

The ward or clinic nurse should initiate the referral, but the coordinating nurse should also collaborate to prevent a breakdown of continuing nursing care, or worse, conflicting advice. It has been our own experience and that of others that most hospital ward nurses, because they frequently have limited time and experience outside a hospital ward, are less able to adequately initiate or coordinate referrals. [5,6] Previous experience in public health nursing is a definite asset for the coordinator who should be familiar with community facilities, health agency policies, and home evaluations. [1,2] Dr. Louise Smith , [6,2] indicates that,on the whole nurses who have had experience in public health nursing make the best referrals for home nursing.

Prompt transmittal of the referral to the public health agency is essential. If the patient is hospitalized, the initial telephone contact with the health agency should be made by the nurse responsible for coordinating the referrals on or before the day the patient is discharged. Written information then follows within 24 hours, with a copy for the family physician and a second for the patient's clinical record. A follow-up request from a clinic visit is handled in a similar manner, depending upon theneeds of the patient and his family.

A lag in the referral makes the public health nurse's work more difficult and the patient often suffers needlessly. (Bladder irrigations were ordered b.i.d. for M.V. with an indwelling catheter. Although the mother was instructed how to do this in the hospital, she felt insecure and did not perform the procedure. The catheter plugged, and the patient had to be admitted to the hospital because of acute urinary tract infection). Because a patient or his family has demonstrated efficiency carrying out a procedure in the hospital, there is no reason to judge him capable of duplicate independent performance in the home. Reinforcement by the nurse of learned processes or procedures is so important in assuring continuity of care.

Follow-up Mechanism: The date the referral is sent to the agency should be noted in a recall file and reviewed by the coordinator. Failures of agencies to supply progress reports on a referred patient should be investigated, as these reports are important indices of continuity of care. In most instances, the public health nurses' progress reports on patients are sent to the nurse coordinator. She should be responsible for providing channels by which these reports can be reviewed by appropriate members of the health team. Thereby, the coordinator in her communication position can be responsible for keeping hospital, clinical staff and community informed of patients current status. Simultaneously, she will be aware of alterations in the health facilities available in the community and in the referral health center. Such direct involvement assists the coordinator in her teaching capacity.

Teaching: The nurse coordinator also functions as a resource person, nursing care consultant, model for student nurses, and teacher of self-care skills to patients and their families. Much informal teaching can take place at patient care orientated conferences.

According to Windmuth,[4] the real test of medical, nursing and social scientific advance lies in the understanding which the public has of it, and its ability to utilize this knowledge effectively in daily life. As a resource person, the nurse coordinator should be well informed about her hospital, the multiple community services available, and the advances and changes in science and technology. She should be able to provide community persons with knowledge about the referral center services in addition to new trends in medical care and nursing care. In turn, she can provide referral center personnel with changes in available local community patient care resources.

Nursing care consultation available in the referral center may be accomplished in written form or by telephone or person-to-person colloquy. The best is , according to our experience, person-to-person colloquy centered around a child and his family of interest to hospital, clinic group, and community agency representatives. Such colloquy best takes the form of a patient conference which can be either in the hospital or community. Our conferences are of three types: (1) patient-care oriented conferences in the University Center, (2) patient-care oriented conferences as part of inservice education in the community, and (3) patient-care oriented conferences as part of formal education. Whatever method of contact is used, the nurse should have an opportunity to discuss nursing care, patient needs, and problems related to the ramifications of a particular disease process which involves the child, family, and community. The interpretation of an individual patient's needs provides the ideal circumstances for teaching both students and colleagues. The opportunity to ask and answer questions increases both community and center nurses' knowledge and understanding of problems and better satisfies the needs of the agency and community.

It should be kept in mind when providing nursing consultation that the subject matter should be contemporary, of proven value, and designed to meet the greater variety of needs represented by each disease, child, family, and community. Consideration should be given to limitations of time, personnel, and facilities present in most community agencies.

To apply or to recommend new knowledge and concepts of patient care at the referral center and community level, the coordinator must actively continue nursing and medical education through postgraduate courses, seminars, conferences and workshops at the local, regional, and national levels. She should constantly review current nursing and medical literature. A part of informal self education is continuing contact through patient care with other members of specialty groups such as psychology, orthopedics, urology, medical-social work, pediatrics, and etc.

Research: In addition to formal professional education, the nurse needs freedom for research. This type of position must provide time opportunities to develop new and innovative methods and techniques of patient care. Collaborative studies are a natural development in the academic setting in which the nurse coordinator is functioning. [7,8] Individual nursing projects with medical consultation can also be developed. [9]

The Public Health Nurse

So far in our discussion, the primary focus has been on the function of the nurse coordinator in the clinical setting. What then are the functions of the public health nurse in the community? Her importance as a member of the health team can never be underestimated. It is through her functioning that patient care is continued in the home.

The simple fact of the matter is that our program, and I am sure many other like programs, are crucially dependent upon public health nursing. This is the reason: we formulate theories about why these spina bifida children and their families are like they are. We bring them into the hospital or clinic and lay out a very neat and orderly program in order to start moving the child from a life characterized by passive inertia to one of active particpation. We spend time working with the parents.

To my knowledge, every major failure in our program has occurred because the provision for continued care in the home has been neglected or not followed through with. The P.H.N. is often the pivotal figure around which the treatment and progress of many of these children and their families turn. She may be the one professional who has access and understanding of all the community resources available for care.

A continual family education and counseling program is a vital aspect in the management of these children and their parents. The program must be flexible to meet the ever changing needs of the parents, child, siblings and relatives. All members of the team are involved in this. However, the continued long-term reinforcement of the learned processes so essential for these families has to be assumed by the public health nurse. Her continued nursing diagnosis of the needs and her nursing action are crucial to realistic management of these multi-problem families.

If the patient is of school age and attending school, it may be more feasible for the school nurse to share in the responsibility of continued care. At such times, close cooperation is required between the school nurse and the public health nurse. The school nurse may be responsible to the nurse coordinator in the clinic in:

1. providing information as to the child's adjustment to school, which may have medical, self-care, social or emotional aspects
2. providing nursing care or supervision of others who have accepted the responsibility of giving care
3. attending clinic conferences to share in planning, organizing and evaluating of a continuous health care plan
4. interpreting the clinic recommendations to the various school personnel concerned with the child

SUMMARY

With the increasing emphasis on early hospital discharges and home care programs, the nurse members of the health team should assume responsibility for coordinating activities associated with implementation of the medical program and for the provision for continuing high quality nursing care in the hospital, clinic and patient's home. As nursing and medical research described new care; as older practices are discarded; as organizations develop new programs and delete others; and as personnel and policy change, an ongoing interagency method of providing continuity of patient care is essential.

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THE ADOLESCENT WITH SPINA BIFIDA

Ida Nakashima, M.D.

Until recently few children born with spina bifida reached the age of adolescence. The statistics were overwhelmingly against such odds. According to Laurence, only about 35% of the cases that reach a large medical center survive until the age of twelve years. [1] Of the 51 children who are seen in our Spina Bifida Clinic, only 6 have attained this age and the majority are severely handicapped in mind and body.

The happy reasurring progress of normal infancy and childhood are denied to children born with such a life-threatening defect. Multiple operations and hospitalizations, prolonged separations from family, all take their physical and emotional toll. Early operation to close the defect, perhaps a ventricular shunt to prevent hydrocephalus, deprive the baby of early mothering care. When the baby comes home, the mother, often struggling with her own feelings of guilt and grief, and sometimes overwhelmed by the responsibility of caring for a fragile and damaged child, is unable to respond in a warm and accepting manner. [2] Such mothers are often afraid of their babies, and fear that holding and handling the infants will injure them.

Such a child, too, since he is partially or completely disabled in his lower extremities, can be deprived of the stimulation and satisfaction of crawling and walking and exploring his environment. Control over his bowel and bladder can never be achieved in the normal manner.

Because his condition necessitates so many visits to the doctor and to specialty clinics for his multi-problem condition, he is also likely to have his sense of dependency and helplessness accentuated.

Schooling is a more difficult experience for him because he has had fewer opportunities to meet new and unexpected situations. Other children look upon him as having lower status than themselves and the handicapped child is more likely to have conflicts with "normal children", i.e., those not handicapped. [2] Not only does he have to struggle with the negative reactions of the people he meets, but he also has to contend with his own feelings of inferiority, of being different in an undesirable way. He is a very special kind of minority group.

Having then survived adverse statistics, operations, hospitalizations and exposure to some kind of education, the child with spina bifida reaches adolescence, with its convulsive strivings for independence, its search for self-identity and clarification of sexual identity.

However, because many adolescents with spina bifida are mentally retarded, these problems of maturation are frequently delayed until fifteen or sixteen, rather than beginning at a normal twelve or thirteen. Furthermore, because these children have had to be so dependent on others, both emotionally and physically, they often do not begin to experience these urgings and strivings until mid to late teens.

In the normal course of growth, the child develops a concept of himself as an individual and as a person, his own body-image; "the way his body appears to himself." [3] During adolescence, with its rapid bodily changes, the adolescent must drastically reconstruct this body-image of himself, further stimulating anxiety, self-consciousness and feelings of inadequacy in the normal adolescent. [4]

The adolescent with spina bifida struggles with these same feelings but in addition, he has a real and undeniable handicap. At an age when "belonging" is so important, the feeling that he is not like others because of his defect produces a feeling of inferiority and loss of self-esteem. Indeed, so strong is this feeling that in adolescents with malignant disease, their immediate concern when informed of their diagnosis is not "Will I die?", but, "How will this disease make me different?" [5]

His peers, who are trying to cope with their own tenuous self-concepts, tend to accord highly discriminatory treatment to persons with physical handicaps. They, too, are threatened by the idea of disfigurement and weakness, and often defend themselves against such feelings by treating handicapped peers as those of lower status, frequently ostracizing them, often treating them with open contempt and hostility. [4] The adolescent with spina bifida may be so totally concerned about himself, and how others make him feel, that it is impossible for him to appreciate the problem his normal peers are having. Interaction between them can be especially painful at this time. [5]

Defenses the Handicapped Adolescent Can Use

Often the handicapped adolescent reacts with anger and depression at the weakness represented by his disability, and at the injustice of his fate. [2] Sometimes such a person seems to go through a mourning process, at last relinquishing the physically intact person he can never be. Often the anger is projected toward those closest to him in his family, and may include those identified with the management of his illness. [3] The adolescent may become sullen and hostile, miss appointments, refuse to communicate, forget to take his medicines and become a difficult management problem.

Others may cover their depression with a Pollyanna attitude which serves as an impenetrable wall, hiding their real despair behind a facade of submissive compliance and passivity, but often with an underlying depression.

Some adolescents may cope by intellectualizing, which helps to master anxiety produced by their condition, and enables them to isolate the feeling from the thought, repress the feeling, and place emphasis on the rational aspect of the knowledge. Such patients are extremely learned about their disease and can discuss it with great facility.

Because so many of the handicapped person's satisfactions must be derived vicariously rather than through direct involvement, they may become sensitive observers, and develop a rich fantasy life to compensate for this lack of activity and social involvement. [2] The intellectually bright ones may pursue academic learning with great fervor to make up for physical lacks.

Dependence - Independence

Adolescents normally have a difficult time relating to adults because of their ambivalence regarding their drive for independence while still having many dependency needs, particularly in early adolescence. This can be intensified in handicapped adolescents who may interpret as weakness the dependent role which their disabilities force upon them and they may resent their families and the physician for making them appear weak. [5] This feeling is reinforced by the many hospitalizations and operations they may have to undergo.

Sexual Identification

Since handicapped children are brought up with the same expectation of being a boy or being a girl and since these children do not usually have ambiguous genitalia, their sexual identification should be firm. In terms of being a sexual person, however, competing with normal rivals for the attention of a member of the opposite sex, the handicapped adolescent feels himself hopelessly outclassed.

For both girls and boys, the prospects of a happy close relationship with a member of the opposite sex seems gloomy. Realistically, having a handicap of this kind does cut down the number of prospective partners to say nothing of sexual difficulties involved.

For boys, particularly the future appears dismal for prospects of marriage. Since many boys with spina bifida have suffered damage to those nerve segments from L-1 or L-2 to S-4, all of which need to be intact to achieve erection and ejaculation, a normal sex life appears to be out of the question, but there are no studies available to corroborate this. We do not know whether such boys are fertile or not, neither do we know how fertile the females with spina bifida are, nor what risk they carry for bearing children with defects of the central nervous system. Furthermore, those more severely affected are totally anesthetic to a level below L-1 or L-2. We don't know what kind of sexual sensations they have because, unlike the paraplegic, they have never experienced normal sensations.

While much has been written about the sexual problems of those with traumatic cord injuries or with cerebral palsy, there is almost nothing in the literature concerning this problem in adults with spina bifida. This appears to be an area which has totally escaped the concern of physicians and psychiatrists alike, partly because so few children with spina bifida have survived to adulthood. Now, with improved medical and surgical care in early infancy and childhood and better medical management of urinary tract infections, many more of them will reach the adult years. We need to know more about their sexual and emotional needs as adults so that we can find ways to help prepare them for these kinds of problems.

Importance of Family

The importance of the child's family in his adjustment to the stresses of adolescence cannot be over emphasized.

" A child born with a congenital defect has to face not only the crippling anatomical and physical defects of such a handicap, but also the emotional reaction of his family and of society - a reaction which may be even more crippling to his total emotional and physical growth than the physical defect itself." [3]

Much of the way a person feels about himself comes from his perception of other people's perception of him. If the people about a handicapped child react with repugnance or abhorrence, the child will come to regard his deformity in the same way. His body image, to a large part, is due to the pleasure or displeasure emotionally significant people find in the child's appearance. Children who are accepted by their families neither over-evaluate nor under-evaluate their bodies. [4] As the body image is strong and acceptable, so a child's total personality is strengthened.

If the family is a stable one and has been able, perhaps with help, to accept the patient's condition and has developed a warm and accepting relationship with him then they, and he too, have a far better chance of surviving the stresses and strains of adolescence.

Physician's Role

The doctor as the primary physician, knows the patient and his family the best, and has helped him through medical difficulties in the past. He sees him as a whole person and is in the best position to coordinate his care. Many patients, because of the complex nature of their multiple handicaps, receive their care through large clinics, and their care tends to be scattered amongst various specialists.

Indeed, a syndrome of "adolescent deterioration" has been mentioned in those chronically handicapped children with cerebral palsy, who share many problems in common with the patient who has spina bifida. There seems to be a worsening of their physical disabilities, often accompanied by depression and a hopeless attitude, perhaps because of a new awareness of their disability.

In our Spina Bifida Clinic, a special effort is made to keep the pediatrician as the primary physician. He sees the patient first, treats him for his cold or his urinary tract infection, follows his progress and tries to be available to him between scheduled clinic visits.

The management of the adolescent slump involves careful repeated counseling of patient and the parents and the continued attentive interest of a responsible physician.

The adolescent, despite a temporary estrangement from his parents, needs a trustworthy, dependable adult who listens to him and promotes a relaxed non-judgmental atmosphere. Those problems which relate to sex especially, might well be handled by the doctor, who can help the adolescent understand his own feelings. The success of therapy depends in part on how well the adolescent can trust his doctor, and how well he relates to him because, unlike the treatment of children, the care of the adolescent involves direct confrontation.

The use of one-to-one relationship with the adolescent, keeping the parents in the background, can encourage the adolescent to take more responsibility for his own illness through his sense of involvement and participation in his own treatment program. Hopefully, he can be helped to achieve a more mature attitude toward his condition so that he can be weaned from frequent therapy to greater self-care and responsibllity.

Working closely with the adolescent, however, does not preclude working with the parents as well. The parent's attitude toward the handicap, their anxieties and fears, their unrealistic fantasies about it must be dealt with. Here, social workers are of invaluable assistance, since they can obtain much family background and give parents the feeling that they have someone for themselves to whom they can turn when they feel the need for supportive help. Sometimes, seeing the parents and adolescent together around specific issues can help clarify misunderstandings around a particular treatment program.

The doctor must help the adolescent to accept the realistic limitations his condition imposes upon him, and to free him from unrealistic restrictions which the adolescent's own anxieties have constructed. He must somehow convey a positive and optomistic approach, pointing out to the patient what kinds of future goals are attainable, and that a gratifying and useful adulthood may still be open to him. [7]

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SUMMARY

Arthur Robinson, M.D.

We have in the last two days heard a great deal about a serious, common (3/1000), complicated congenital malformation which obviously requires the highest quality of comprehensive, inter-disciplinary, and coordinated care. No simple or easy answers have been given. If they were simple, this symposium would not have been needed.

One characteristic of the meeting which perhaps has been striking is the note of optimism which has pervaded it. This is in marked contrast to what has appeared in the literature and what one often senses in house officers, nurses and practitioners with only a passing exposure to the disease. This is of the utmost importance at a time when a society with innumerable calls on its resources is trying desperately to define its priorities.

Our task as health professionals, as I see it, is in its broadest aspects a simple one. It is our job, when we embark on the therapy of these patients, to do all in our power to make them the best persons they are capable of becoming. Dr. Sharrard sounded this note when he stated that to treat these patients in any way but aggressively is to maim them.

The most effective way, of course, to treat a disease is to prevent it. In a disease with unknown cause, this is difficult, except perhaps by the use of genetic counseling, and this I think should be offered to all families with this condition.

Although much attention, and rightly so, was paid to the more or less engineering aspects of repairing the structural and locomotor problems (and most impressive among these were Dr. Sharrard's insistence on repair within 12 hours) , a gratifying amount of time was spent on the effect of the child's interaction with his environment on his development. To this end, we heard about the importance of the infant's early "conversation" with his mother, to borrow a phrase from Spitz, in his ultimate development of awareness of his perceptual problems and his self-image. His interaction with his parents, both father and mother, will affect him at least as much as all children and possibly more. Parental attitudes, we heard, will be affected by society's and medicine's interest in the special strain this disease imposes on the family unit, although Dr. Sharrard has several times made the important point that we may overstress the likelihood of problems arising in given families. The therapeutic effort of parent and parent groups has only been touched on but I would like to stress again their importance.

Pervading the entire approach is a true feeling of sympathy for the individuals and families concerned. I stress the word true since, to quote Lord Lister, "When you adopt sympathy as a principal, you abandon it as an emotion."